Sacrococcygeal tumours and cysts in adults

Charles Court, Gilles Missenard

Spine and Tumor Unit, Orthopaedic and Traumatology Department
Bicêtre University Hospital, Paris-Sud University

Cite: Court, C, Missenard, G. Sacrococcygeal tumours and cysts in adults. In: Maigne JY and Doursounian L (eds) Coccyx Disorders. Proceedings of the First International Symposium on Coccyx Disorders, Paris, 2016. pp 57-66.

SACROCOCCYGEAL TUMOURS AND CYSTS IN ADULTS

Charles Court, Gilles Missenard
Spine and Tumor Unit, Orthopaedic and Traumatology Department
Bicêtre University Hospital, Paris-Sud University

INTRODUCTION

Primary bone tumors are rare and localization in the sacrum is even more infrequent. Tumors in this area have several histological diagnoses. Tumor can starts from the sacrum and spread in the surrounding tissue or starts in the surrounding tissues and eventually invade the sacrum. The diagnosis is often delayed. This delay can lead to huge tumor volume which rends the surgical treatment more challenging. The more volume the tumor is the more sequellae after surgical resection the patients have. In dealing with bone tumors one must consider always to do a biopsy before starting any treatment. The biopsy must follow guidelines to avoid contamination and spreading of the tumor.

Treatments include chemotherapy, radiotherapy and surgery depending of the histological diagnosis. Strategy of the treatment is decided in a multidisciplinary team including orthopaedic oncologist surgeons, chemotherapist, radiotherapist, and radiologist. In this presentation we will present the diagnosis and treatment of sacro-coccygeal tumors.

DIAGNOSIS

Clinical presentation

Tumors of the sacrum are often delayed diagnosis. Clinical symptoms are poor and unspecific at beginning. Coccigodynia can be present or pelvic discomfort or bowel constipation. Pain can have inflammatory type in case of malignant tumors with permanent pain more and more resistant to pain killers. Due to the poor clinical symptoms tumors are discovered when the volume is important and responsible of intractable pelvic pain or neurological symptoms such as cauda equina syndrome. Around the sacrum there is plenty dead space and no fragile structure which can explain that sacral tumor can have huge volume before symptoms. Sacral tumors are rare entity and medical doctor are not aware and consequently don't look for. The more common diagnosis error is with low back pain or incomplete sciatica, gynecologic or colorectal pathologies. In case of low back pain MRI or CT scan can be performed but it is not uncommon that radiologist center the exam on the lumbar spine and do not include the entire sacrum which

can miss distal tumor of the sacrum. Similarly gynecologist will focused on genital area and the Ultrasound examination will missed the sacral area. Finally, the diagnosis can be made before a coccyx injection for pain, usually the radiologist will do an MRI.

To summarize, the diagnosis of sacral tumor is delayed, leading to huge tumor volume. The clinical presentation is unspecific and one must consider doing an MRI including the entire sacrum in a patient with coccydynia - or lumbar pain with a normal lumbar spine on MRI - especially with inflammatory pain and before any coccygeal injection. [Fig 1]

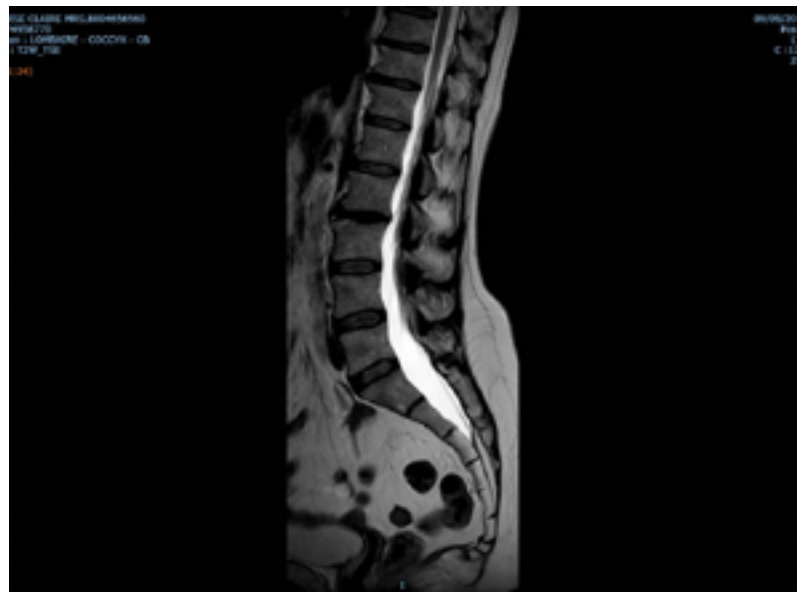

Fig 1: MRI including the sacrum and the coccyx. The distal chordoma is located at the button limits of the field of this MRI, but visible in this case

Radiologic diagnosis

X-rays of the sacrum are often performed at first. If the tumor is not responsible for lytic lesion of the bone the x-rays remain normal. The main exam is the MRI to diagnose tumors and its extension in the soft tissue especially the relationship with the rectum and the invasion of sacral bone and the sacral nerve root. TDM is mandatory to explore if the tumor is aggressive to the bone in examining if there is or not a scalloping or a bony condensation at the junction of the tumor with the bone. In case of bony condensation this means a slow tumor development inversely if bony destruction is important the tumor is an aggressive lesion spreading rapidly.

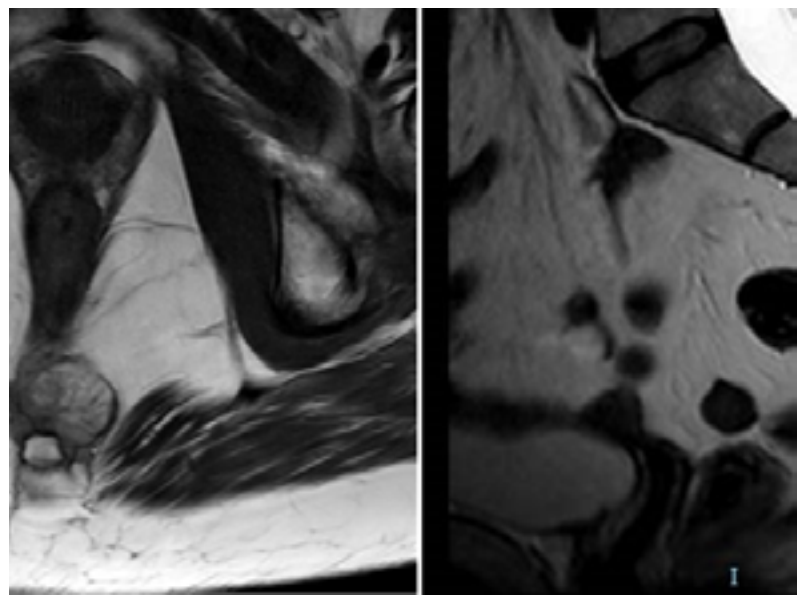

Fig 2A, 2B: Axial and sagittal views on MRI showing a small tumor of the coccyx, needle Biopsy revealed the diagnosis of chordoma

Extension work up needs a total body scan looking for other localization, and a full spine MRI searching for other spine lesion.

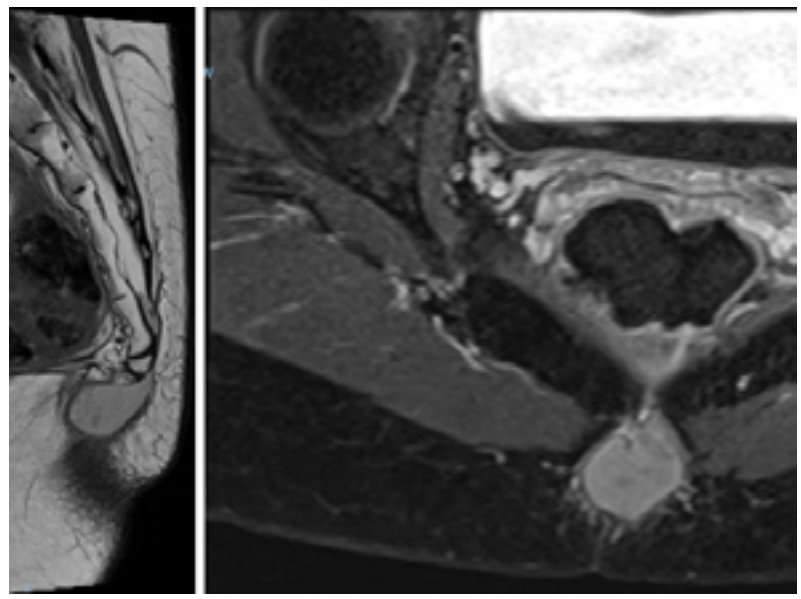

Fig 3A, 3B: Axial and sagittal views on MRI showing a small tumor of the coccyx, Biopsy revealed a dermoid benign tumor

Biological diagnosis

Standard blood cell count is mandatory with PCR and VS looking for infection.

Biopsy

In dealing with tumor a biopsy is always mandatory to have a histological diagnosis to plan the treatment strategy. Imaging can orientate the diagnosis but never gives the right diagnosis [Fig 2A-B] [Fig 3A-B]. Biopsy must follow the general rules of soft tissues or bone tumors treatment strategy (ref). Biopsy must be performed in a reference center to avoid wrong biopsy and histological misdiagnosis.

Study have shown a decreased in survival rate if the biopsy is not performed in a reference center able to treat sarcomas. There is no indication for a so called "biopsy-excision" which leads to an "unplanned surgery" or a "oops-surgery" with poorer outcome [16]

A needle biopsy can be performed under local anesthesia guided with Ultrasound or CT Scan. The Biopsy strategy must be decided with radiologist and oncologic surgeon. The biopsy tact must be direct to the tumor, on the way of the future surgical access for the resection. The biopsy tract must never go through the spinal canal if not invaded, neither trough the rectum or through the peritoneum. If these structures are invaded by the biopsy they will need to be resected which will make the surgical "en bloc" resection more difficult or impossible leading to a poorer survival rate [16,18]

Surgical biopsy can be performed with the same rules if more tissue is necessary for histological analysis or in case of non-contributive needle biopsy, which can occur in 20 to 30% of cases.

In some cases such as a cystic lesion in the retrorectal area, biopsy may be not technically possible because the approach will contaminate the surrounding tissues. More than this, it will spread out the tumor and the risk of infection is high. In some special circumstances and after discussion in a multidisciplinary team a surgical excision with wide margins if possible can be performed.

HISTOLOGICAL DIAGNOSIS

Sacro-coccygeal tumors can be benign or malignant and only the biopsy can confirm the histological diagnosis nor the radiological appearance or "doctor feeling" or experience. One must remember that tumor of the sacrum and coccyx are more likely to be malignant than benign.

Malignant tumors

Bone metastasis

After 4O years of age, bone metastasis is the first diagnosis to consider regarding frequency. If there is no recent history of cancer a biopsy will be performed. Hematologic primary tumor such as Lymphoma and Myeloma must be ruled as well.

Chordomas

It is a low grade primary malignant bone tumor originating from embryologic notochord. The incidence is of 0.5 per million. Tumor usually occurs in the 4th and 5th decades but can develop earlier even in childhood. Fifty percent of chordomas develop in the sacrum, 40% at the cranio-cervical junction (childhood expression) and 10 % in the lumbar spine. Histological examination reveals typical physaliphorous cells. Brachyurie gene expression is present in 8% of cases. The differential diagnostic can be difficult with benign notochordal cell tumors [1,9,10,14]. Radiologycally chordomas are bone lytic lesion, with extraosseous extension. Benign notochordal cell tumors usually showed homogeneous hyperintensity on T2-weighted images with no enhancement with gadolinium. Usually chordomas have heterogeneous signal and enhancement with gadolinium[Fig 4-6].

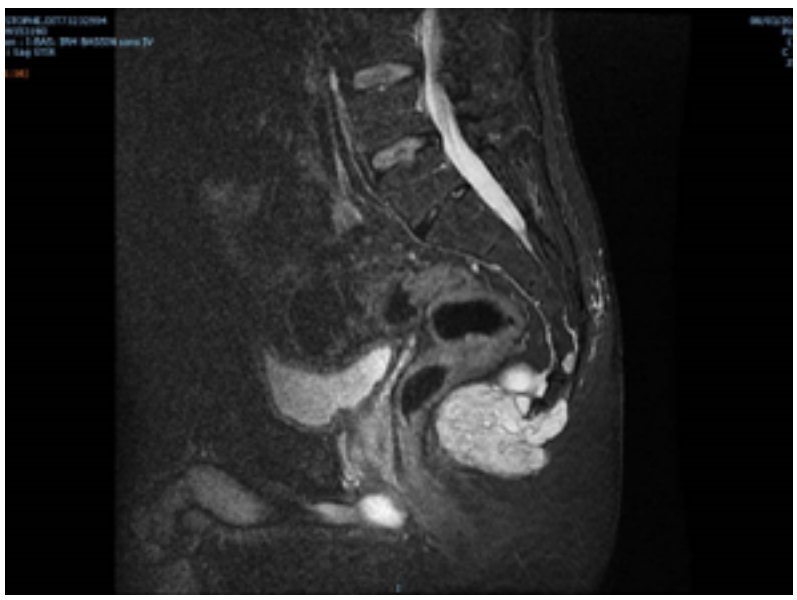

Fig 4: T2-weighted MRI showing hyperintensity of coccyx chodorma with an anterior development toward the rectum

On biopsy the differentiation can be impossible and one should consider treating as a chordoma an aggressive tumor with bone lysis, high contrast enhancement. Benign notochordal cell tumors can be capable of malignant transformation. Chordomas can lead to bone or lung metastasis but it is mainly a local aggressive tumor with a high rate of recurrence even at long term [2,4,18].

Rectal leiomyosarcoma

Rectal leiomyosarcoma is an infrequent tumor in less than 0.1% of cases [7]. Most of them grow in the lower third part of rectum and men are particularly affected. Extension of these tumors is mainly local, by contiguity. They can invade rectal mucosa leading to ulceration and bleeding. Computerized tomographic examination reveals a heterogeneous tumor, well-defined, multilobular, with cystic necrotic components.

Other malignant tumors

See further the malignant ganglioneuroblastomas, malignant Schwanommas or Neurofibroma

Benign tumors

Pilonidal Sinus or pilonidal cyst **[Fig 7-8]**

It is the most frequent lesion in the sacrococcygeal aera. 1% of male and 0.1% of women are affected. The predisposing factor is still unknown; aetiology can be an ingrown hair in the gluteal cleft, pressure in prolonged sitting position (truck driver), obesity are predisposing factors [5]. Most of the time people are asymptomatic and diagnosis is made in emergency because of pain and infection necessitating a surgical excision.

Teratomas

Teratomas are congenital tumors diagnosed in the childhood (90% of cases diagnosis is made before the age of 5) or even in fetal and diagnosed during pregnancy [6,11]. The incidence is evaluated to be 1 teratoma every 35 000 or 40 000 birth. Geminal cells can be localized in extragonadique aeras such as the presacral area, coccyx or mediastinum. Approximately 20% of sacrococcygeal teratomas are identified prenatally; 70% are identified at birth, and the remaining 10% are identified within the first year of life and usually present with intestinal obstruction [20]. It is the most frequent presacral tumor in childhood. Rarely these tumors are revealed in the adult age (ref simpson dis colon).

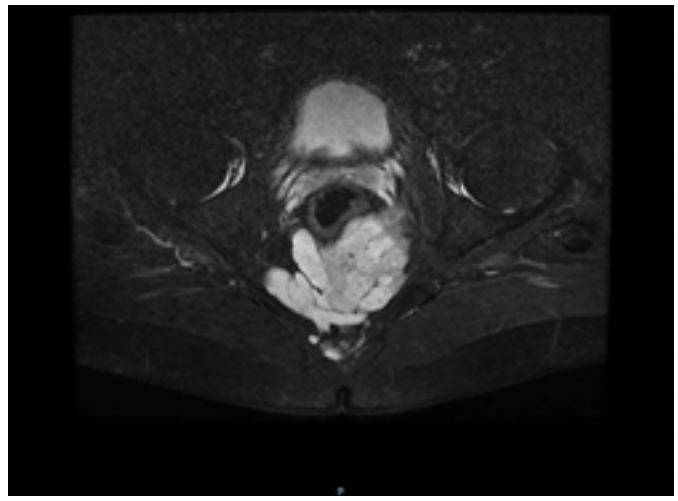

Fig 5 : T2-weighted MRI showing hyperintensity and heterogeneity of coccyx chodorma polylobular with an anterior development around the posterior part of the rectum

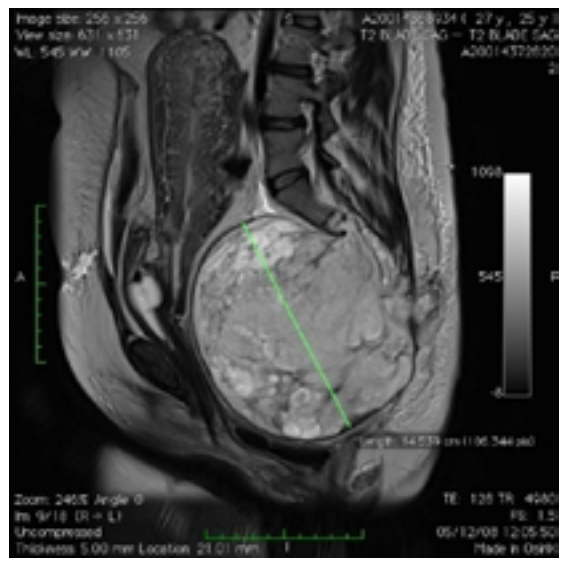

Fig 6 : T2-weighted MRI showing hyperintensity and heterogeneity of coccyx chodorma with an anterior development and destruction of the sacro-coccygeal area till S1. Tumor can be asymptomatic during a long time before diagnosis

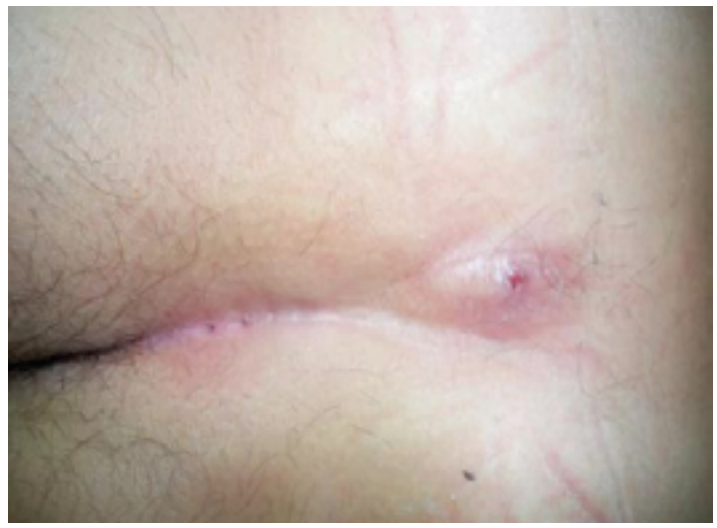

Fig 7 : Pilonidal cyst presenting as a median posterior tumor located more proximal than the coccyx

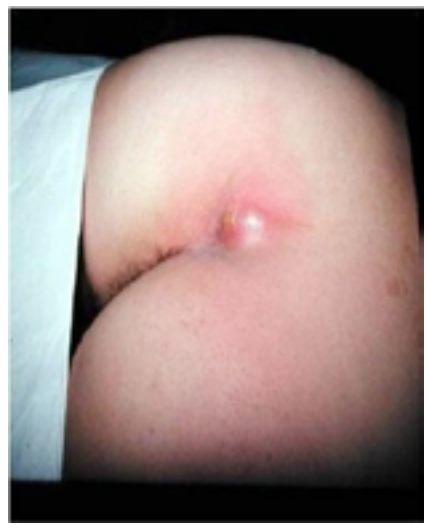

Fig 8: Infected Pilonidal cyst presenting as a median posterior tumor

For this author among 26 patients during 33 years, 5 were malignant. The radiological presentation was a cystic lesion or mixed solid (with fat or calcification) and cystic. This dys-embryonic tumor is Benign but can have a late recurrence (ref), it is a rare diagnosis in adulthood.

Retroperitoneal pseudomixom

It is a rare clinical entity characterized by excessive mucin secretion. Glandular epithelium of the cyst is responsible for mucin secretion. Nuclear tomography and magnetic resonance sections formation show a homogeneous, well-defined, cystic, septated. Frequently mural nodules or calcifications are observed in the cyst.

Retro rectal lesion is very rare entity most of them are embryonic cyst are the most frequent epidermoid, dermoid [Fig 9], enteric or neuroenteric cysts [7,13]. They are asymptomatic or revealed by chronic infection with fistula or pelvic abcess. Anal gland cysts are mucus-secreting cysts. Magnetic resonance examination reveals a retrorectal uni- or multi-locular cystic lesion, near the anal sphincter.

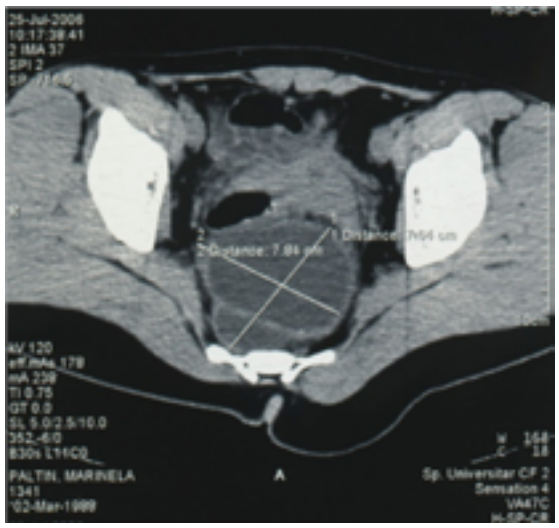

Fig 9: Retrorectal dermoid cyst

Bone tumors

The sacrum is the third most common location for giant cell tumors after the knee and the radius. Giant cell tumors typically affect patients in the second to fourth decades, with a female predominance. Sacral giant cell tumors usually develop in an eccentric position, but commonly extend to involve both sides of the midline. There is usually a thin cortical rim, but the soft tissue mass can break through this rim; the mass is usually large before it is detected and may extend into the presacral space. Although generally classified as a benign tumor, 5% to 10% of giant cell tumors have been reported to be malignant. Many of the malignant lesions are thought to be related to previous radiation therapy. Furthermore, giant cell tumors occasionally metastasize to the lung although the primary lesion is considered histologically benign. Localization starting at the coccyx is very rare: it is usually a tumor extension from the sacrum.

Other bone sarcomas

Ewing's sarcomas, chondrosarcomas can affect the sacrum and extend to the coccyx but are very rarely starting from the coccyx

Tumor from neurologic tissue

Tarlov cyst

Also known as perineural, or sacral nerve root cysts. They are dilations of the nerve root sheaths and are abnormal sacs filled with cerebrospinal fluid (CSF). They are located most frequently at the S2, S3 level of the sacrum [Fig 10-11].

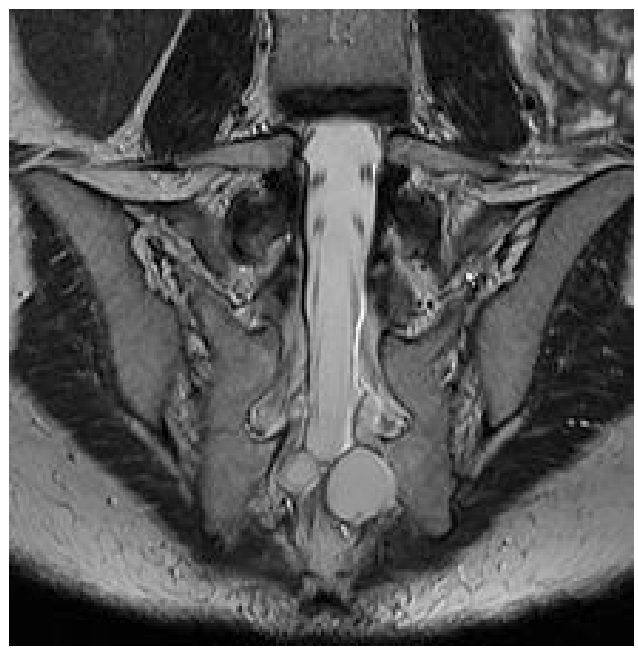

Fig 10: Tarlov Cyst on coronal view on MRI imaging

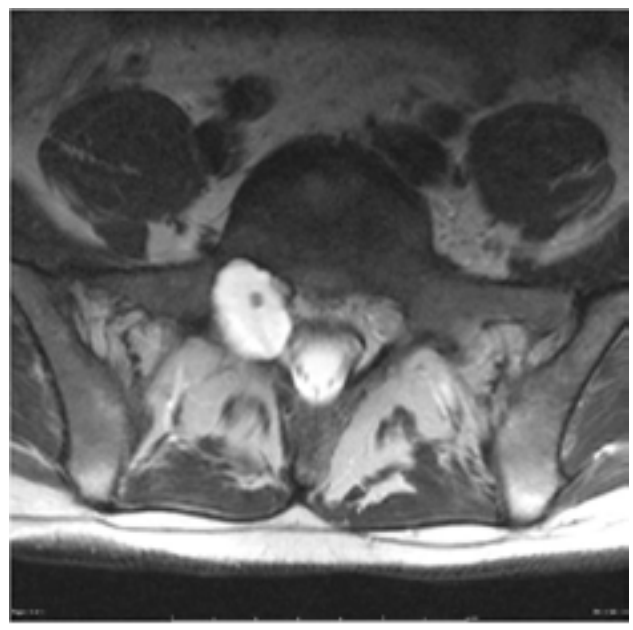

Fig 11: Tarlov Cyst on axial view on MRI imaging with bone scalloping

These lesions have been estimated to affect between 4.6 and 9% of the adult population more frequently in women. Tree types are described: Type I or «extradural meningeal cysts without spinal nerve root fibers»; Type II or «extradural meningeal cysts with spinal nerve root fibers» (that is real Tarlov cysts); and Type III or «spinal intradural meningeal cysts.» The hydrostatic and pulsatile forces of CSF cause Tarlov cysts to grow over time which can cause bone scalloping [Fig 12]. Responsibility of Tarlov Cyst in radicular pain, or paresthesia is very difficult to confirm. As the mass enlarges, sensory nerve root filaments are stretched over the periphery of the lesion or are compressed against adjacent bone or other nerve roots, causing pain or other sensory disturbances. Approximately one fifth of Tarlov cysts greater than 1.5 cm can be symptomatic, and can produce, according to their anatomical location, a variety of symptoms of nerve root compression, including radicular pain (sacral, coccyx, hip, perineal pain) paresthesias, and urinary or bowel dysfunction.

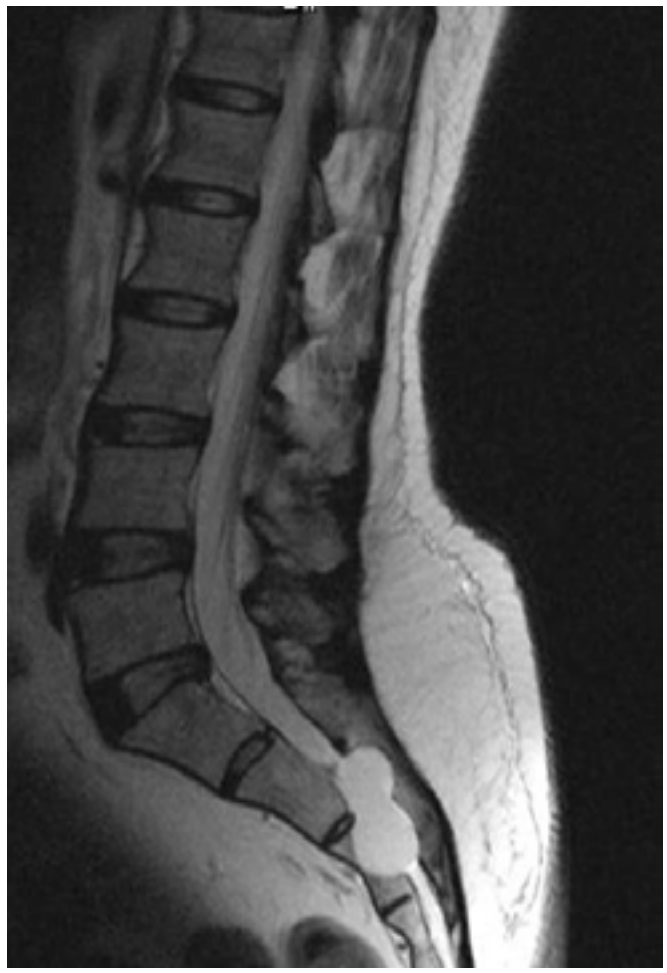

Fig 12: Tarlov Cyst on MRI imaging with bone scalloping

Schwanommas-Neurofibroma [Fig 13-14]

Benign tumor can develop from the sacral nerve. Tumors develop through the neural foramens in toward or outward from the sacral canal and are shaped like a dumbbell [8]. These tumors have a slow development, inward tumor are generally not big due to the defined space of the sacral canal but outward tumors can reach very huge volume before giving any clinical symptoms. Pain is usually the main symptoms with an inflammatory symptomatology, before giving any neurological deficit. Most of the benign neurogenic tumors are shown as a homogenous signal change on MRI, with about 6% showing as a cystic change; however, most malignant neurogenic tumors are shown as an inhomogeneous signal change in the MRI, with about 75% showing as a cystic change. Therefore, the appearance of an inhomogeneous signal and cystic change in huge neurogenic tumors indicates the possibility of a malignant change. An imaged guided biopsy is mandatory especially in case of history of irradiation therapy. In such case, tumor is considered of a high risk of malignant transformation in neurosarcoma.

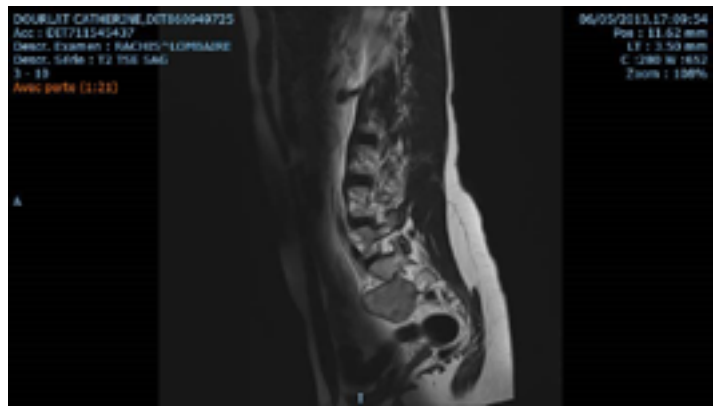

Fig 13: Sagittal view showing the schannoma existing from a foramen

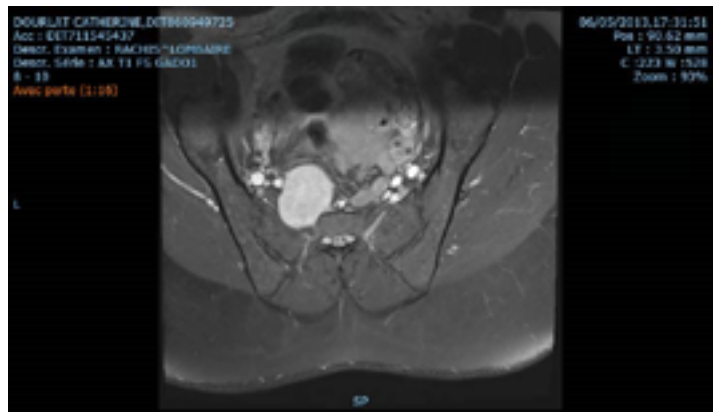

Fig 14: axial view showing the tumor inside the foramina with contrast enhancement with gadolinium

Ganglioneuroma [Fig 15-16]

It is rare entity originating from neural crest and developed in the Sympathetic nerve chain developed from autonomic cells, lying in the presacral aera in 37.5% of cases or in the mediastinum in 41% of cases [12,19]. It can occur in neurofibromatosis type 1. The benign (ganglioneuromas) and malignant (ganglioneuroblastomas) forms of these tumors are virtually identical radiologically. They are asymptomatic and discovered incidentally on radiography. 60 % of cases the diagnosis is before the age of 25 years.

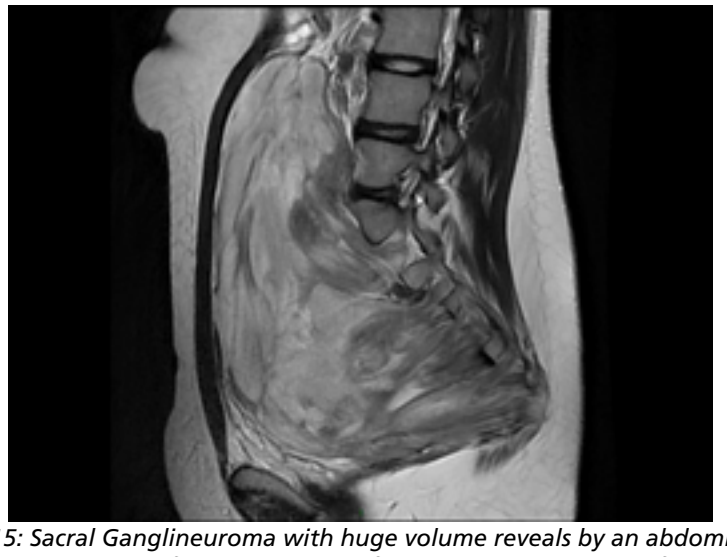

Fig 15: Sacral Ganglineuroma with huge volume reveals by an abdominal palpable mass. Tumor follows the sacral foraminas characteristic of the tract of the sympathetic chain

Ependynoma

Spinal ependymomas most commonly occur as intramedullary tumors throughout the spinal axis. In the lumbosacral region, ependymomas are most commonly associated with the conus medullaris and cauda equina, but can also occur extradurally in the sacrum, presacral tissues, or subcutaneous tissues over the sacrum. Intradural ependymomas, especially those in the lumbosacral region, are now recognized for their potential to spread throughout the central nervous system (CNS), whereas extradural tumors elicit more concern for their association with extraneural metastases.

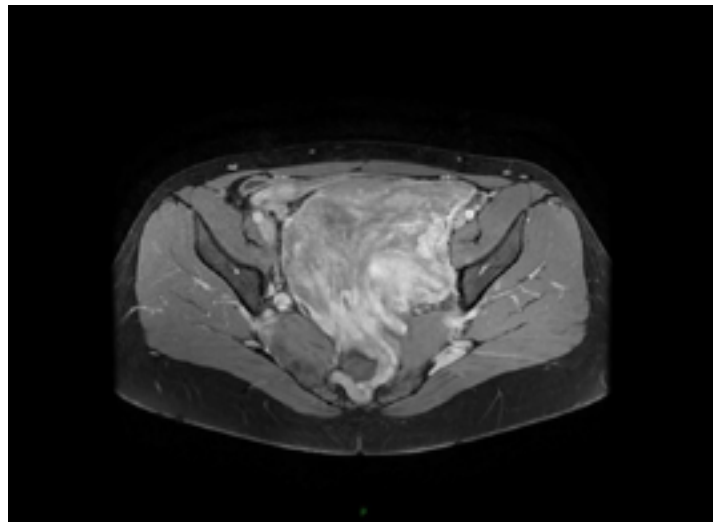

Fig 16: In the axial plane tumor inside the canal getting outside the canal through the foramina. No bone destruction

Sacral ependymomas arise from ependymal cells of the terminal filum, expand the sacral canal, and are usually of the myxopapillary type or metastases from a primary tumor of the central nervous system, causing an erosive mass of the sacrum [Fig 17].

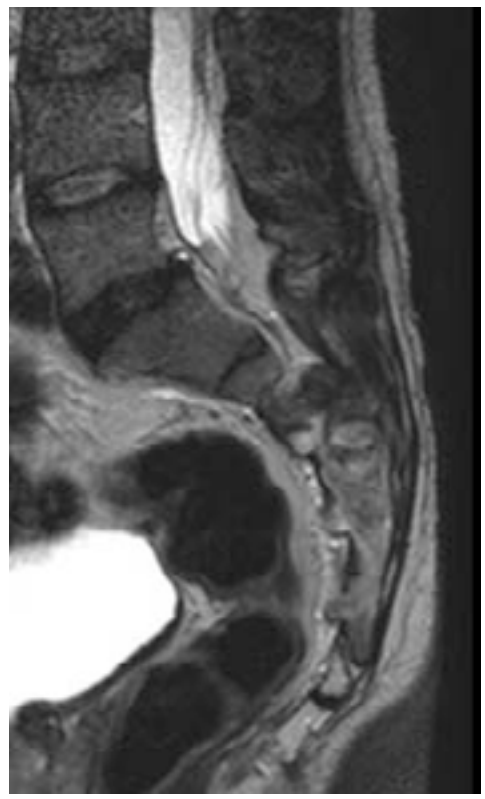

Fig 17: Ependynoma limited inside the sacral canal with extension to the coccyx

Malformation of the sacrum

Anterior sacral meningocele due to anterior agenesia of the sacrum is usually associates with other malformations [15]. Tomography and magnetic nuclear resonance examination, confirm the bone defect and highlights well defined cystic tumor, unilocular, developed in the prerectal space. MRI images can show a communication between meningocele and tecal bag.

TREATMENT

Benign tumors

These tumors are treated by surgical marginal excision. When tumors have little volume neurological deficit or bowel sequellae are infrequent. Recurrence is rare. Some tumors necessitate specific treatment such as Denosumab and embolization in Giant cell tumor before surgical curettage.

Tarlov cyst treatment is very controversial, patients presenting with symptoms exacerbated by postural changes or Valsalva maneuvers are likely to benefit most from surgery. Some authors have proposed corticosteroid perineural injection, TENS therapy, micro-surgical cyst fenestration, along with a fat - or muscle graft reinforced cyst closure. Results are unpredictable and majority of doctors and surgeons does not think that Cyst Tarlov can be responsible for symptoms and does not need any surgical treatment.

Schwanommas-Neurofibroma are treated depending of the extension. Tumor localized inside the spinal canal can be excised from a posterior approach and sacral laminectomy. When tumors spread outside the canal in the anterior space an anterior approach is necessary combined with the posterior approach [8].

Malignant tumors

Treatment is very challenging. Considering the survival rate, surgery needs an “en bloc resection” with wide margins to avoid local recurrence.
Chordomas are resistant to chemotherapy and radiotherapy and “en bloc” resection is the recognized “gold standard” treatment (ref). The prognosis relates to its completeness and the violation of the tumor margins at the initial surgery [18]. Survival rate was 84.4% at 5 and 10 years, survival rate with local recurrence was 64 and 56%, respectively, after 5 and 10 years [3].

When tumors are located distally to S3 foramina a posterior approach can be sufficient to achieve an “en bloc” excision if anterior tumor development is limited. Tumors beyond S3 need an anterior and posterior approach and sacrifice of nerve root if often necessary leading to perineal deficit. Surgical resection consists of an anterior approach for vascular control, retro rectal dissection from presacral space and elevation of an omentum flap. Then patient is positioned on prone position to expose the sacrum, opening of the spinal canal, section of the nerve root when invaded by the tumor. Then an osteotomy of the sacrum is performed to resect the sacrum with the tumor and surrounding tissue without tumor violation. Posterior reconstruction is performed by omentum flap and mesh to avoid posterior rectal herniation. Adjuvant radiation therapy can be performed in some cases [2,3,18].

Complications

Infection is the most frequent complication in sacral tumor related to proximity to the anus. Infection rate increases with complexity and duration of surgery and varies from 10% to 50% [3,18].

Neurological deficit of the perineum is the most impairment for the patient. Bladder, anal and genital dysfunction depends of the nerve root sacrifice. If S3 nerve root can be preserved bladder function can be efficacious. For distal tumors, deficit is very infrequent, and if anal external sphincter can be repair, bowel function will be preserve.

CONCLUSION

Tumors originating from the coccyx are rare, most of them are invasion from a sacral tumor. One must always keep in mind that a coccydynia or pelvic numbness can result from tumor, especially in case of inflammatory pain resistant to pain killers, and one should always look for perineal dysfunction, bladder or bowel symptoms. Malignant tumors such as metastasis and chordomas are the most frequent event; benign tumors are more likely originating from neurologic structures. Teratomas are the most frequent diagnosis in childhood. Pilonidal cyst is usually revealed by infectious complication.

MRI including the entire sacrum and coccyx is mandatory especially in case of incomplete sciatica without diagnosis on lumbar MRI. Discovering of a tumor needs to refer patient to a center specialized in orthopaedic-oncology to perform a biopsy and define a strategy in a multisciplanary team.

BIBLIOGRAPHY

1. Amer HZ, Hameed . Intraosseous benign notochordal cell tumor.M.Arch Pathol Lab Med. 2010 Feb; 134(2):283-8
2. Dubory A, Missenard G, Lambert B, Court C. Interest of Laparoscopy for «En Bloc» Resection of Primary Malignant Sacral Tumors by Combined Approach: Comparative Study With Open Median Laparotomy. Spine (Phila Pa 1976). 2015 Oct 1;40(19):1542-52.
3. Dubory A, Missenard G, Lambert B, Court C. «En bloc» resection of sacral chordomas by combined anterior and posterior surgical approach: a monocentric retrospective review about 29 cases. Eur Spine J. 2014 Sep;23(9):1940-8.
4. Dubory A, Missenard G, Court C. Late local recurrence, at 19 and 17years, of sacral chordoma treated by en bloc resection. Orthop Traumatol Surg Res. 2016 Feb;102(1):121-5.
5. Eryilmaz, R., Sahin, M., Okan, I., Alimoglu, O., Somay, A. (2005) Umbilical pilonidal sinus disease: predisposing factors and treatment. World J. Surg.2005; 29: 1158–1160.
6. GatcombeHG,AssikisV,KoobyD, etal. Primary retroperitoneal teratomas : a review of the literature. J Surg Oncol 2004;86:107–113.
7. Glasgow SC, Birnbaum EH, Lowney JK, Fleshman JW, Kodner IJ, Mutch DG, et al. Retrorectal tumors: a diagnostic and therapeutic challenge. Dis Colon Rectum 2005; 48:1581-7.
8. Guo Wei, Tang Xiaodong, Yang Yi, ,Tao Ji. Strategy of Surgical Treatment of Sacral Neurogenic Tumors.Spine 2009;23:2587-2592
9. Haasper C, Länger F, Rosenthal H, Knobloch K, Mössinger E, Krettek C, et al. Coccydynia due to a benign notochordal cell tumor. Spine. 15 juin 2007;32(14):E394-6.
10. Kreshak et al. Difficulty distinguishing benign notochordal cell tumor from chordoma further suggests a link between them. Cancer Imaging 2014, 14:4
11. Lam CH, Nagib MG. Nonteratomatous tumors in the pediatric sacral region. Spine. 2002; 27(11):E284-E287.
12. Modha A. Presacral ganglioneuromas. J neurosurg Spine 2005;2:366-371
13. Munteanu M. Retrorectal dermoid cyst: a rare clinical entity. Current health sciences journal. 2013;3;179-183
14. Nishiguchi T1, Mochizuki K, Ohsawa M, Inoue T, Kageyama K, Suzuki A, Takami T, Miki Y. Differentiating benign notochordal cell tumors from chordomas: radiographic features on MRI, CT, and tomography. AJR Am J Roentgenol. 2011 Mar;196(3):644-50
15. O'Neill OR, Piatt JH Jr, Mitchell P, Roman-Goldstein S. Agenesis and dysgenesis of the sacrum: neurosurgical implications. Pediatr Neurosurg. 1995; 22(1):20-28.
16. Pretell-Mazzini J, Barton MD Jr, Conway SA, Temple HT. Unplanned excision of soft-tissue sarcomas: current concepts for management and prognosis. JBJS Am. 2015;97(7):597-603.

17. Ruggieri P, Angelini A, Pala E, Mercuri M. Infections in surgery of primary tumors of the sacrum. Spine. 2012;37(5):420-8.
18. Ruggieri P, Angelini A, Ussia G, Montalti M, Mercuri M. Surgical margins and local control in resection of sacral chordomas. Clin Orthop. nov 2010;468(11):2939-47.
19. Vardas K and al.Presacral ganglioneuroma :diagnosic consideration and therapeutic strategy.Case rep oncol 2013;6:561-568
20. ZhangC,BerneyDM,HirschMS, etal. Evidence supporting the existence of benign teratomas of the postpubertal testis: aclinical, histopathologic, and molecular genetic analysis of 25cases.Am J Surg Pathol. 2013;37:827–835.