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Occasionally report of sacral chordoma; treatment and review of literature

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SUMMARY: **Occasionally report of sacral chordoma; treatment and review of literature.**

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Introduction. *Brain notochordal cell tumors (BTCN) are lesions arising from notochordal differentiation which affect the axial skeleton.*

Presentation of case. *We report a case of a patient treated in our General Surgery Unit of the University Hospital of Bari, Italy, with occasional finding of sacral chordoma at the histological examination.*

Discussion. *Because of their location, sacral chordomas can affect bowel and bladder with organ specific symptoms. Radiotherapy may be used as a palliative treatment or for recurrence in those patients who cannot be submitted to surgery.*

Conclusions. *Due to the high local recurrence rate radiation therapy should be considered mandatory after any type of chordoma resection. Multidisciplinary management of the disease is mandatory and improve patient outcomes. Patients should have maximal tumor debulking with adjuvant radiotherapy when possible.*

KEY WORDS: Brain notochordal cell tumors - Chordoma surgical treatment.

Introduction

Brain notochordal cell tumors (BTCN) are lesions arising from notochordal differentiation which affect the axial skeleton. The notochord is an embryo mesodermal structure which brings signal tissues for organization and differentiation. With a literature reported prevalence of 20% in autopsy and with an increase of magnetic resonance imaging (MRI) investigation, findings are more frequent. At the beginning, those lesions are characterized by non-specific symptoms until they increase, they can be large, and symptomatic or they can be associated with different and non organ specific symptoms. Chordomas are malignant neoplasms arising from not complete regression of notochordal tissue along the cranio-coccygeal axis; they are low-grade, slow-growing but locally invasive and locally aggressive tumors and they belong to the sarcoma family. These neoplasms arise from nothocordal remnants cells and occur in the midline along the spinal axis from the clivus to the sacrum, before the spinal cord. The distribution of chordomas localization is 50% in the sacral region, 35% in the skull base and 15% occurs in the vertebral bodies of the mobile spine (most commonly on C2 vertebrae followed by lumbar and thoracic spine). Because of their location, sacral chordomas can affect bowel and bladder with organ specific symptoms. The diagnosis of this disease in the starting period may be difficult because the patients have non-specific symptoms especially when this tumor is less than 3 cm wide. Overall 5-year survival is approximately 50%; the literature suggests that best treatment is en bloc surgical resection followed by high-dose conformal radiation therapy such as proton beam radiation.

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Case report

We report a case of a patient treated in our General Surgery Unit of the University Hospital of Bari, Italy, with occasional finding of chordoma at the histological examination. A 74-year-old male patient underwent a previous surgical consultation many years before for a non-specific low back pain and for a wrong diagnosis of obturator hernia. During last 6 months the patient complained about a mass extending from the coccyx to the skin of the left gluteus; it was not mobile over the muscular plane, and the patient referred increase of the constipation with frequent paresthesia of the inferior limbs. MRI showed a neoplasm (10x15 cm) occupying the left perirectal area, with dislocation of the rectum and the anal canal on the opposite side and the prostate with a lobulated pseudocapsula with haemorragic area inside; another neoplasm of the left gluteus was reported with the same characteristics (Figures 1, 2, 3). After the MRI, the patient was admitted to our department for a transcutaneous biopsy of the mass, but the pathological examination was negative for tumor cells. Two weeks later the patient underwent a surgical procedure to remove the mass. After spinal anesthesia, the patient was placed in prone position and after incision of the skin a bloody tissue with jelly aspect was found, hardly connected to the coccygeal vertebrae. It was performed a huge excisional biopsy and the specimen was sent for examination. The pathological examination revealed a lobulated tumor composed of groups of cells separated by fibrous septa; the cells have small round nuclei and abundant vacuolated cytoplasm associated with chordoma. The patient was then evaluated by neurosurgeons, radiotherapists and oncologists for the treatment. He is now performing radiotherapy with decrease of the neurological symptoms and with improvement of bowel function with decrease of the constipation and reduction of the mass about 30%. Written informed consent was obtained.

Discussion

Chordoma was first described by Luschka in Virchow's laboratory in 1857 (1). It arises from notochord tissue and has a low growth and occurs along the cranio-coccygeal axis. Although it is slowly growing, chordoma is still defined as a malignant tumor because of its local aggressiveness and metastasis. About 50% of chordomas occurs in the sacrococ-

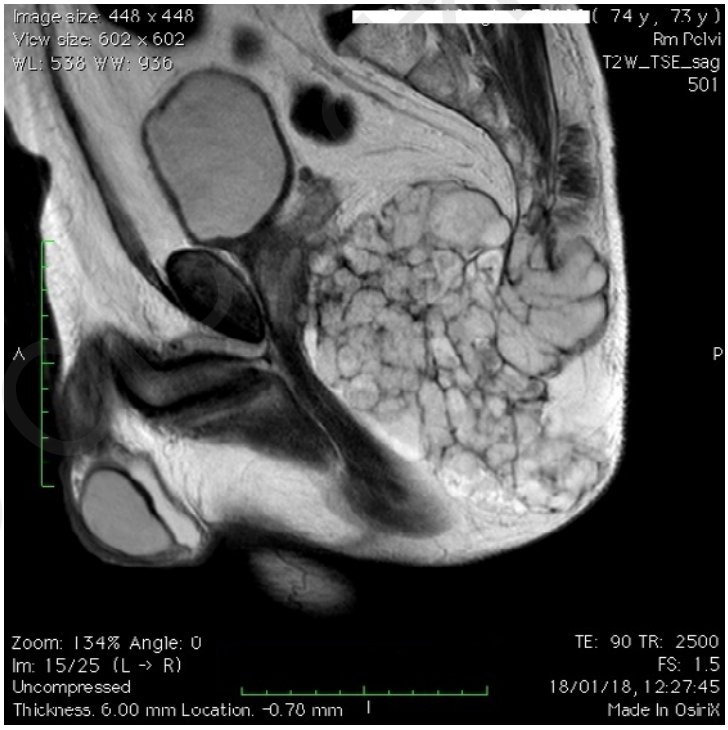

Figure 1 - Sagittal plane of pelvic MRI reporting the sacral chordoma with dislocation of the rectum and the anal canal.

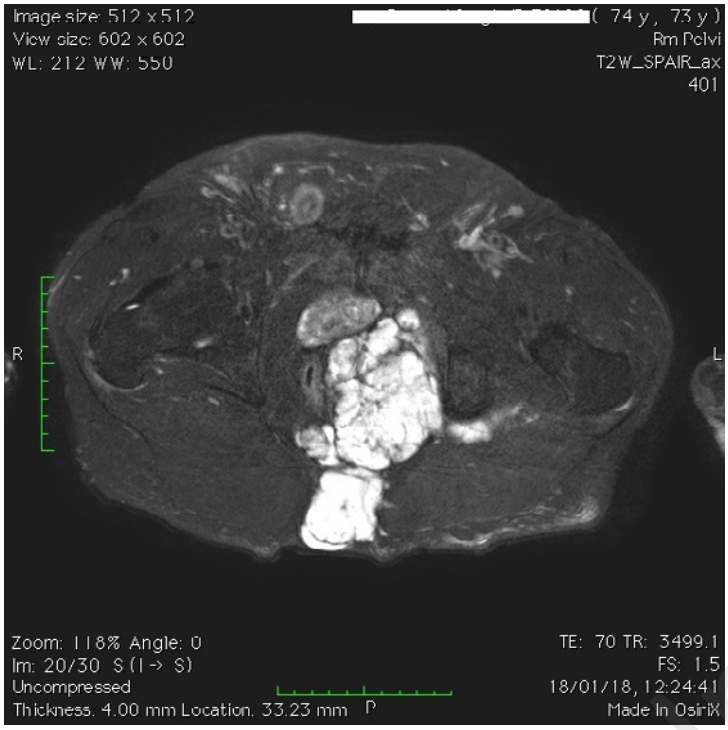

Figure 2 - Axial plane of pelvic MRI of sacral chordoma with evidence of lobulated pseudocapsula.

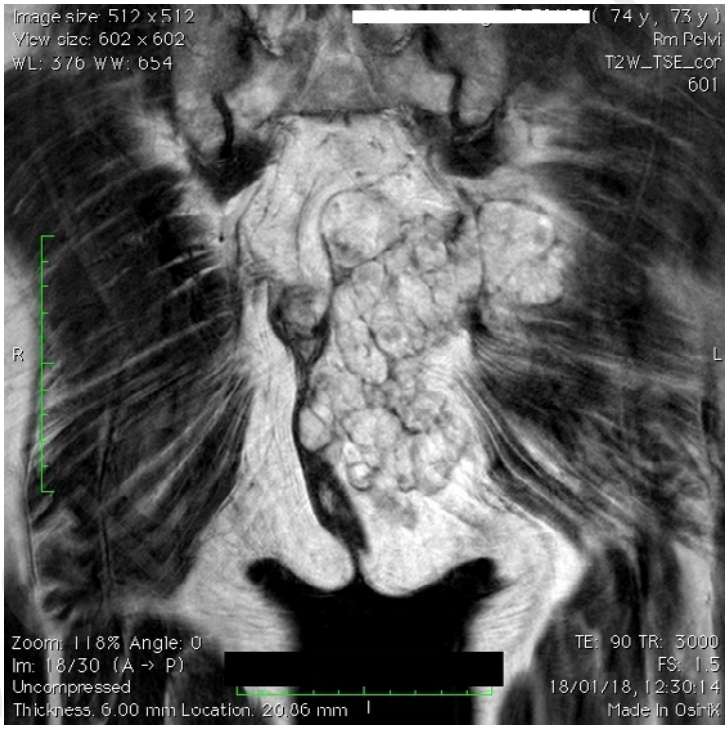

Figure 3 - Coronal plane of pelvic MRI of sacral chordoma with dislocation of the rectum and the anal canal.

cygeal region, 35% in the spheno-occipital region, and 15% in the spine (2). Some reports suggested that BNCT may be the precursor lesion of chordoma (3-5). It is typical in males of 40/60 years with a prevalence of one case on 100,000 persons (6). Symptoms are non-specific and different and are

strictly due to the location of chordoma. For example, patients with clival chordomas may present with headache, diplopia, or impairment of other cranial nerves, while those with sacral chordomas may present with low back or buttocks pain, neuropathy, and/or gait disturbance and urinary symptoms.

Unfortunately, due to the slow-growing nature of chordoma, which results in insidious, non-specific symptoms, diagnosis may be delayed for months or years. The median overall survival (OS) from the time of diagnosis has been estimated at around 6 to 7 years. Macroscopically it may appear as a jelly tumor with some fibrous tissue with blood inside (7). These neoplasms may be classified in 4 different patterns depending from the differentiation grade:

1) classic, the most frequent
2) poorly differentiated, very often in childhood and associated with absence of the gene INI-1
3) not differentiated, extremely aggressive in children
4) chondroid, that is very hard to differentiate from chondrosarcoma.

Himmunochemistry is mandatory in the diagnosis of chordoma with the positive results of the citocheratine, and protein S100 (8). In 2007 Dobashi et al. observed the activation of EGFR (9); in 2013 Zhang (10) et al. confirmed that and referred an association with the MET that is a protein responsible for cellular proliferation and for invasiveness (11). Many other Authors referred an activation of PDGFR and PI3K/aKT/mTOR (12). The gold standard for the treatment is surgery with complete excision trying to remove the tumor and to decrease the risk of recurrence or dissemination of the disease. If not possible a local debulking may decrease symptoms secondary to mass effect and provide a smaller target volume for radiation therapy (13). Radiotherapy may be used as a palliative treatment or for recurrence in those patients who cannot be submitted to surgery. Due to the high local recurrence rate radiation therapy should be considered mandatory after any type of chordoma resection. Despite the low-grade status of chordomas they have a high recurrence rate and significant mortality (14). Five-year overall survival is approximately 50% but improved with complete resection with negative margins to 65% 5-year survival rate. The 5-year survival rate in surgical resection with positive margins is approximately 50% and if the chordoma is inoperable is approximately 40% (15). The slow-growing nature of this tumors makes them resistant to conventional chemotherapeutic agents. Chemioterapy with imatinib and sorafenib shows important results in large tumors decreasing the speed of growth, while the use of corticosteroid may influence the pain arising from nervous compression (16). Recent studies proposed new local therapies as the cryoablation and RFA to improve pain control. Local microwave hyperthermia and high-intensity focused ultrasound (HIFU) may also offer benefit in a palliative setting; however, currently there are no published data supporting their use (17). Most physicians recommend life-long surveillance with MRI. There are insufficient data to recommend exact follow-up for patients with recurrent chordoma. Experts recommend that MRI should be performed every 3-6 months at least for the first 3 years from treatment of LR/local progression (18).

Conclusions

Chordoma management is fairly well defined. Multidisciplinary management of the disease is mandatory and improve patient outcomes. Patients should have maximal tumor debulking with adjuvant radiotherapy when possible. To date, there are no series available in literature combining surgery and intraoperative radiotherapy (IORT). Additional therapeutic strategies continue to emerge and will improve patient outcome like immunotherapy, immune checkpoint inhibitors or brachytherapy.

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