

CASE REPORT

Extraspinal Sacrococcygeal Ependymoma

ABSTRACT

Ependymomas usually arise within the spinal cord or in the brain. Ependymomas originating in the sacrum or extraspinal parasacral soft tissues are rare tumors. A case reported in the subcutaneous tissue posterior to the sacrococcygeal region. Radiologic and pathologic studies are presented. Differential diagnosis and the pathogenesis of extraspinal ependymomas are discussed.

Key Words : Ependymoma, lower extremity, sacrococcygeal region

Introduction

Ependymal tumors usually arise within the spinal cord or in the brain. Spinal ependymomas arise from ependymal cells lining the central canal or its remnants and from the cells of the ventriculus terminalis in the filum terminale. Several different cell types have been described.

The cellular ependymoma is predominately an infratentorial tumor that occurs mainly in children and adolescents. It typically fills the fourth ventricle.

Myxopapillary ependymomas occur exclusively in the spinal cord and are typically found in the conus medullaris or filum terminale. The majority of sacral ependymomas are of the myxopapillary type. These tumors may reach very large sizes. Most myxopapillary ependymomas are slow-growing neoplasms, although some sacral and presacral lesions behave aggressively and metastasize to the lymph nodes, lungs and bone (1).

Subependymomas are small nodular or lobulated at the caudal fourth ventricle or foramen of Monro (2,3).

Ependymal cells may be found within the coccygeal ligament as well as in heterotopic positions so ependymomas may rarely occur in extraspinal locations. Although spinal ependymomas are most often seen in the cauda equina less than 5% of them occur extradurally in the sacrococcygeal region (4,5).

Case Report

A 55 year old man who was operated on because of the cystic lesion posterior of the sacrococcygeal region about 20 years ago, admitted to our hospital because of the presence of a slow growing mass in the intergluteal fold for two years. On physical examination a round, nontender, totally mobil mass, approximately 10 cm diameter, was discovered in the subcutaneous tissue posterior to the sacrococcygeal location. He was otherwise healthy. Both hematological and biochemical blood tests were normal.

Ultrasonography revealed a well defined cystic lesion (85x60 mm in diameter) which contain multiseptal and with solid component next to the wall (Fig 1).

Computed tomography (CT) scan demonstrated 85x60 mm in diameter, at 9 HU density hypodens mass posterior of the sacrum, in subcutaneous fatty tissue (Fig2).

A magnetic resonance imaging study showed the cystic nature of the lesion, with hyperintensity on T2/Weighted images and hypointensity on T1/Weighted images (Fig 3a-b). Especially on superior of the cystic lesion was found nonhomogenous intensity.

At operation, the cystic lesion was found in the subcutaneous soft tissue, unattached to the sacrum and with no connection the spinal canal. Complete local excision was performed.

On pathological examination, macroscopically the tumor measured 9x5x4 cm with a well defined nodular surface. The cut surface was mainly cystic and focally papillary solid. Microscopic examination of tumor tissue revealed round to oval neoplastic cells that were focally arranged in papillary structures with fibrovascular cores. Nucleoid background substance was present in the cores, surrounding vessels in small cystic spaces and there were large areas of necrosis (Fig 4).

Immunoperoxidase studies on formalin-fixed, paraffin embedded tumor tissue showed positive staining for wimentin and glial fibrillary acidic protein and lack of staining for cytokeratins, S-100 protein and neuron specific enolase. The light microscopic and immunohistochemical data were considered diagnostic of myxopapillary ependymoma.

Fig. 1 : Ultrasonography shows well defined cystic lesion (85x60 mm in diameter) which contain multiseptal and with solid component next to the wall.

Fig.2 : CT scan of the pelvis shows 85x60 mm in diameter, at 9 HU density hypodens mass posterior of the sacrum, in subcutaneous fatty tissue.

Fig. 3a-b : MR images show the cystic nature of the lesion, with hyperintensity on T2/Weighted images and hypointensity on T1/Weighted images and nonhomogenous intensity at superior of the cystic lesion.

Fig. 4 : The true ependymal rosettes contain a well defined central lumen, necrosis and blue-tinged musin microcysts

Discussion

Primary extraspinal ependymomas are rare and usually located in the subcutaneous sacrococcygeal or presacral region. The first case was reported by Mallory and about 75 cases since then. It has been reported that 50 of them are subcutaneous variants(6,7).

The differential diagnosis includes sacrococcygeal teratoma, pilonidal cyst, and neurogenic tumor. Sacrococcygeal teratomas are either cystic and solid or predominantly cystic; they are rarely solid over 50% have calcification or ossification. Sacrococcygeal myxopapillary ependymoma may be mistaken for a solid teratoma; however, most of the sacrococcygeal teratomas are diagnosed in the infantile period and imaging studies help indicate the diagnosis, especially if fatty areas are present in the lesion (8).

While it is universally accepted that CNS ependymomas develop from the ependymal cells lining in the ventricles and the central canal, the pathogenesis of extraspinal ependymomas is still debated, with three main hypotheses reported in the literature (9,10,11). The first hypothesis suggests that they arise from the coccygeal medullary vestige, that is an ependymal lined cavity forming from the remnants of the caudal portion of neural tube: ependymal rests have often been found in random autopsies (12,13). Furthermore, this hypothesis could explain the high prevalence of the myxopapillary subtype, which typically originates from the filum terminale. Another hypothesis is that these tumors develop from ectopic ependymal cells originating from the filum terminale, supported by the correlation between neural arch defects and the onset of extraspinal ependymomas. Other authors suggest that they may originate from primordial germ cells with neuroectodermal differentiation.

Following full or near-complete resection, Ependymomas have a good prognosis, with greater than 10 years of survival. Subcutaneous sacrococcygeal ependymomas still show substantial rates of recurrence after a long remission and tend to do distant metastasis (18).

And postoperatively, long-term follow-up should be needed for evaluation of metastasis. We mustn't forget the extraspinal myxopapillary ependymomas can be confused with postsacral mass lesions rarely.

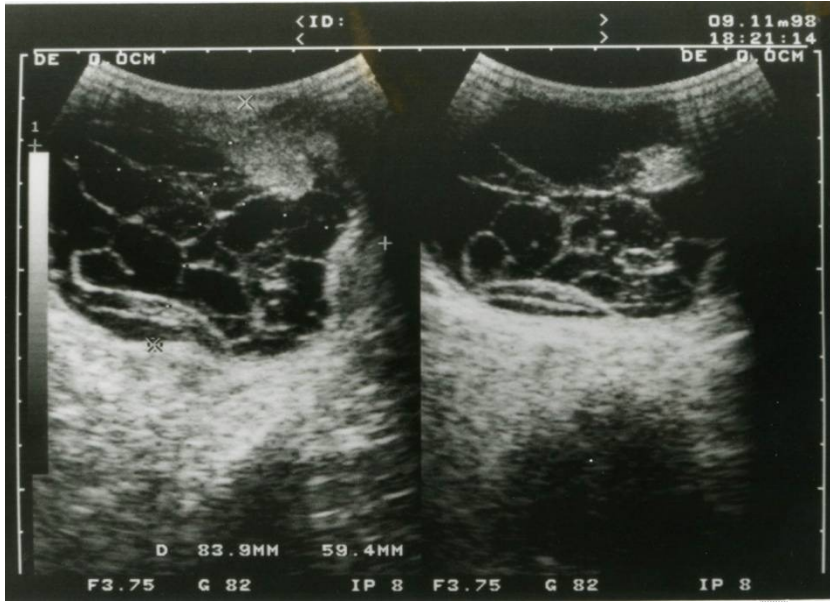


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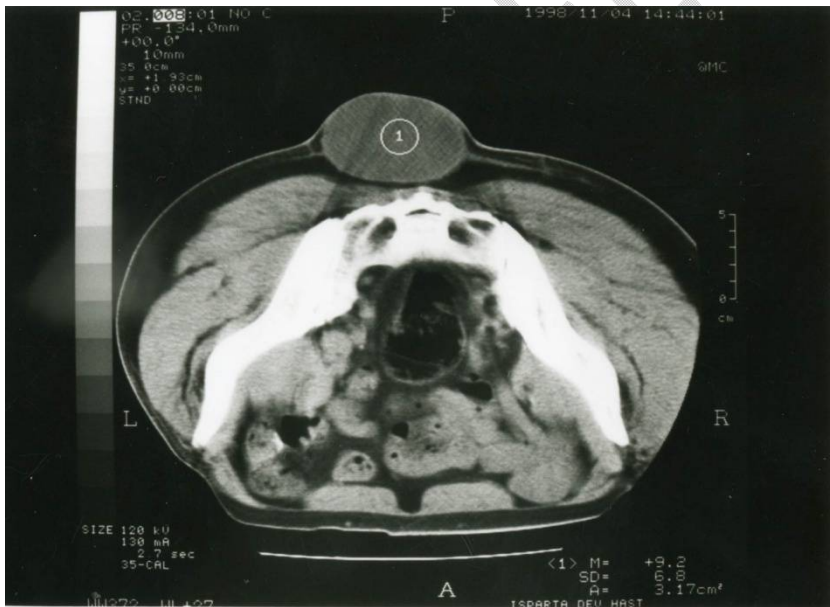


Fig.2 : CTscan of the pelvis shows 85x60 mm in diameter, at 9 HU density hypodens mass posterior of the sacrum, in subcutaneous fatty tissue.

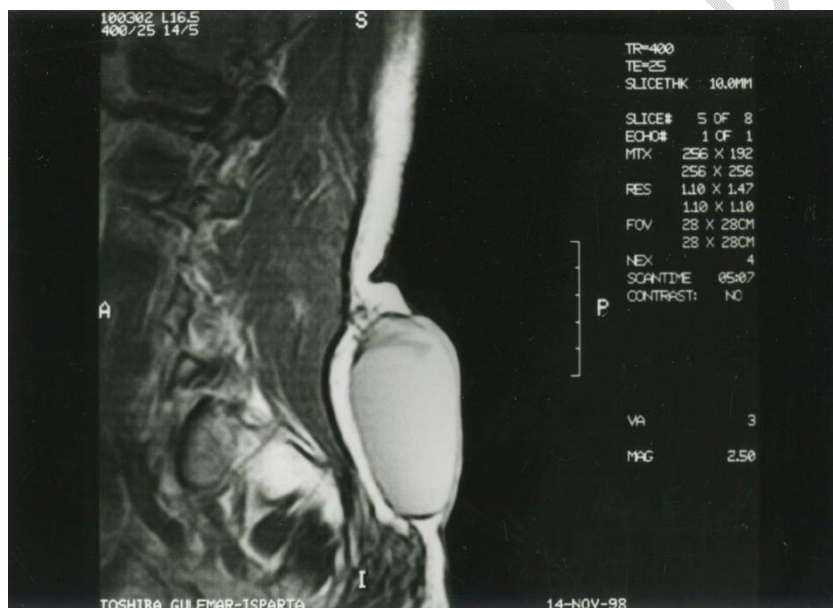
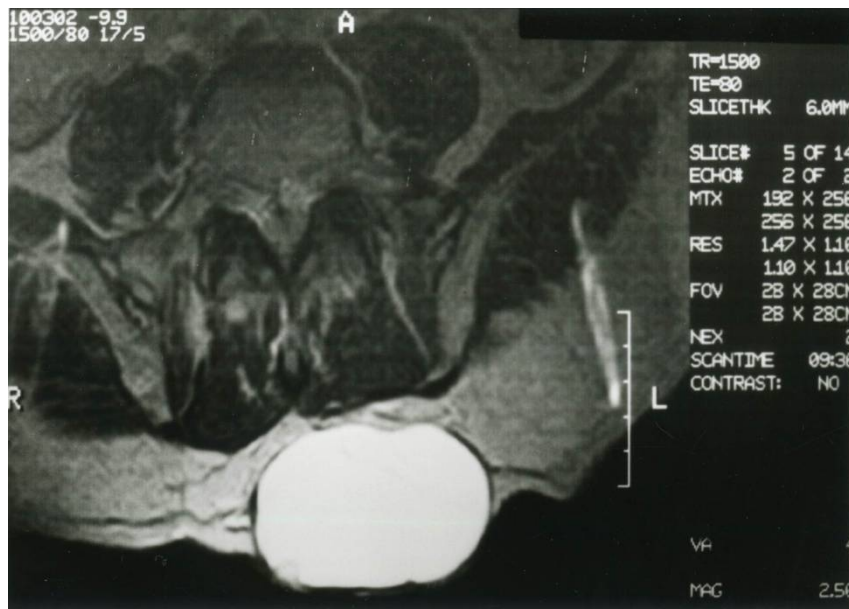


Fig. 3a-b : MR images show the cystic nature of the lesion , with hyperintensity on T2/Weighted images and hypointensity on T1/Weighted images and nonhomogenous intensity at superior of the cystic lesion.

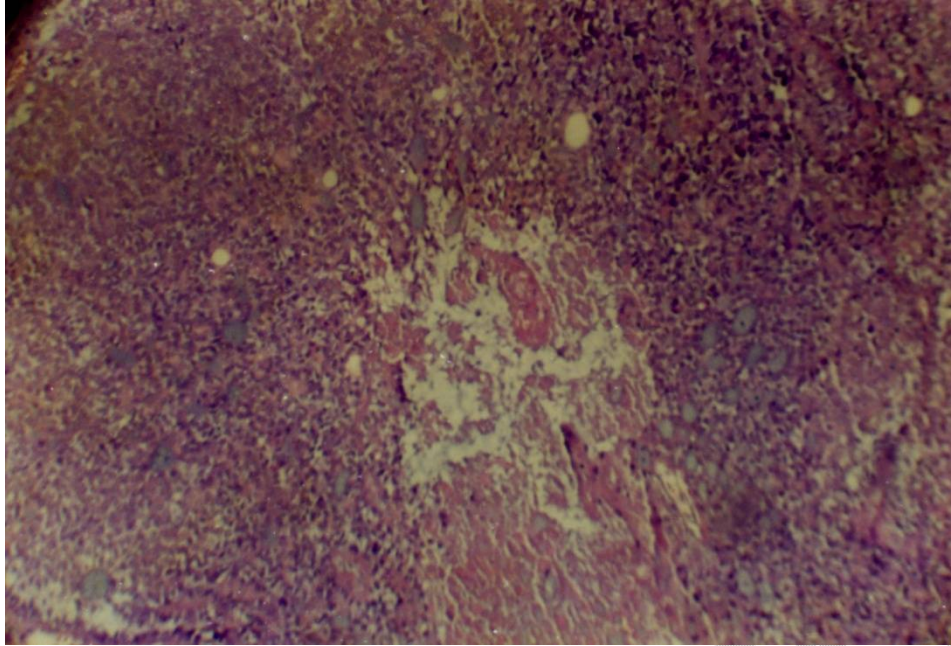


Fig. 4 : The true ependymal rosettes contain a well defined central lumen, necrosis and blue-tinged musin microcysts

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