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Editor
Nancy E. Epstein, MD
Clinical Professor of Neurological Surgery, School of Medicine, State U. of NY at Stony Brook

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Case Report

Anaplastic myxopapillary ependymoma of the sacrum: A case report

Abolfazl Rahimizadeh, Zahed Malekmohammadi, Parviz Habibollahzadeh, Walter L. Williamson, Ava Rahimizadeh

Pars Advanced and Minimally Invasive Medical Manners Research Center, Pars Hospital, Iran University of Medical Sciences, Tehran, Iran.

E-mail: *Abolfazl Rahimizadeh - a_rahimizadeh@hotmail.com; Zahed Malekmohammadi - dr.malekmohammadi@gmail.com; Parviz Habibollahzadeh - phabibollahzadeh@gmail.com; Walter L. Williamson - wlw83@hotmail.com; Ava Rahimizadeh - ava.rahimizadeh@hotmail.com

***Corresponding author:**
Abolfazl Rahimizadeh,
Pars Advanced and Minimally Invasive Medical Manners Research Center, Pars Hospital, Iran University of Medical Sciences, Tehran, Iran.

a_rahimizadeh@hotmail.com

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ABSTRACT

Background: Myxopapillary ependymoma (MPE) with anaplastic features is extremely rare. There are very few such case reports in the medical literature.

Case Description: A 23-year-old female presented with lower back pain, and both urinary and fecal dysfunction. The patient underwent gross total surgical excision of the MR documented expansile intrasacral tumor. The histology was compatible with a MPE containing anaplastic features.

Conclusion: The medical literature contains a few comparable cases of subcutaneous sacrococcygeal MPE with anaplastic components. Here, however, we have a young female with an anaplastic intrasacral MPE treated with gross total surgical excision.

Keywords: Adolescent idiopathic scoliosis, Anaplastic ependymoma, Distal add-on, Filum terminale, Myxopapillary ependymoma, Sacral tumors, Sacrum, Urinary incontinence

INTRODUCTION

Most myxopapillary subtypes of ependymomas occur in the sacral region. However, accompanying anaplastic features are quite rare.[5-8] Here, we present a young female with a history of corrective surgery for idiopathic adolescent scoliosis who, 9 years later, developed an intrasacral anaplastic myxopapillary ependymoma (MPE) requiring gross total resection.

CASE REPORT

A 23-year-old female presented with a 1-year history of low back pain and urinary/fecal dysfunction. Originally, her pain was incorrectly attributed to distal "junctional scoliosis" (i.e. sequela of her previous idiopathic scoliosis fusion surgery at age 14). The plain radiographs documented an expansile intrasacral tumor, while the MR scans showed a lesion that was isointense on T1-weighted and inhomogeneous on T2-weigthed images [Figures 1 and 2]. The CT further confirmed expansion of the sacral canal with enlargement of the foramina (i.e. best seen on sagittal and coronal reconstructed CT images) [Figure 3].

Surgery, histology, and clinical course

The patient underwent a sacral laminectomy for gross total tumor removal [Figure 4]. Histology and

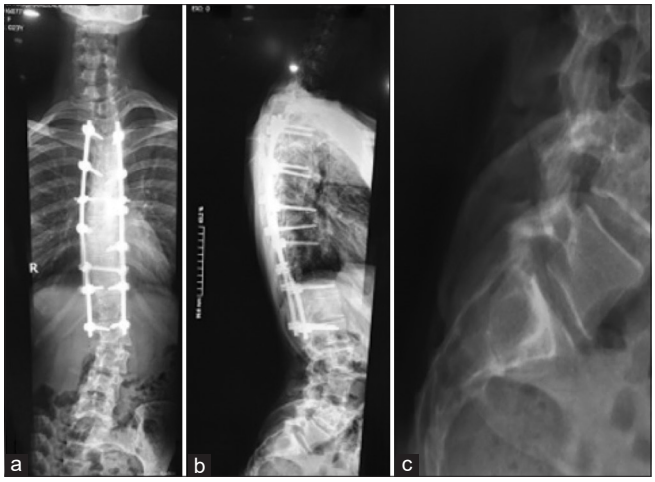

Figure 1: (a and b) AP and lateral radiographs showing thoracolumbar instrumentation. As well as distal junctional deformity. (c) Expansion of the sacrum; (c) localized sacrum shows narrowing and remodeling of the walls of sacrum.

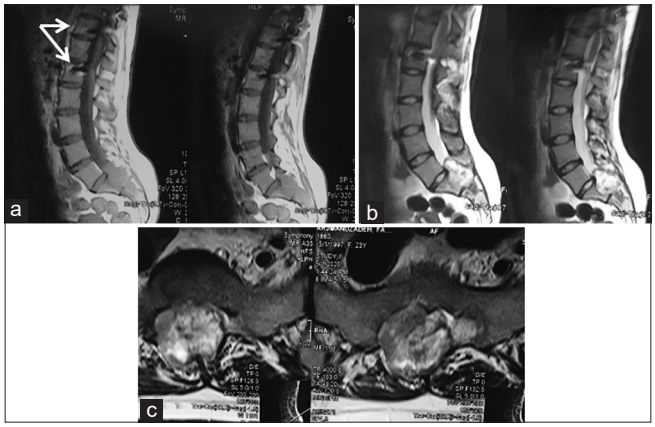

Figure 2: (a) T1-weighted sagittal MRI showing an isointense expansile mass in the sacrum; (b and c) T2-weighted sagittal and axial MRI shows a hyperintense mass with small isointense patches. Screws in L1 and T11 vertebral bodies are seen (white narrow arrows).

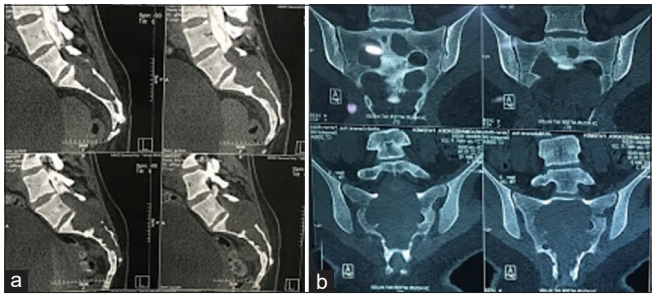

Figure 3: Sacral reconstructed C.T. (a) Sagittal images showing expansion of the sacrum, (b) coronal images note enlargement of the sacral foramina.

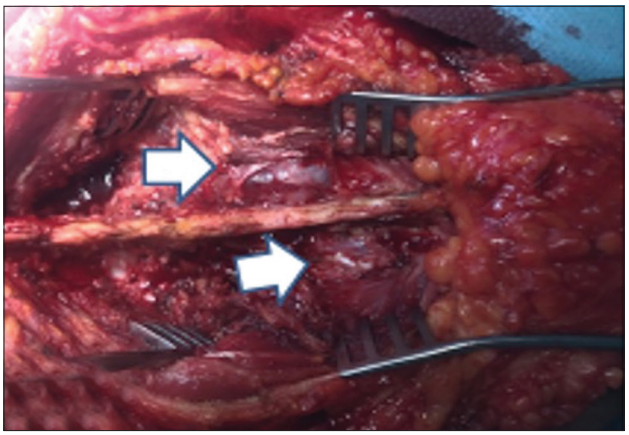

Figure 4: Intraoperative photographs: (a) note protrusion of the tumor from the enlarged sacral foramina (white arrow heads).

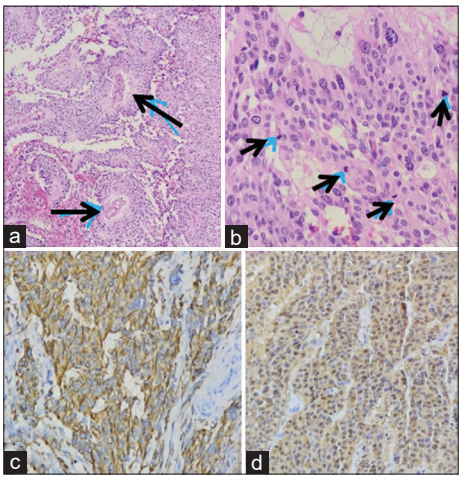

Figure 5: (a) H-E 200 staining shows the cells arranged around vascular core containing blood vessels or pseudorosettes (black arrows), typical for MPE. (b) H-E higher magnification shows cells with anaplasia (black arrow heads), (c) note the tumor cells are diffusely reactive with glial fibrillary acidic protein, and (d) diffusely positive for S100 staining.

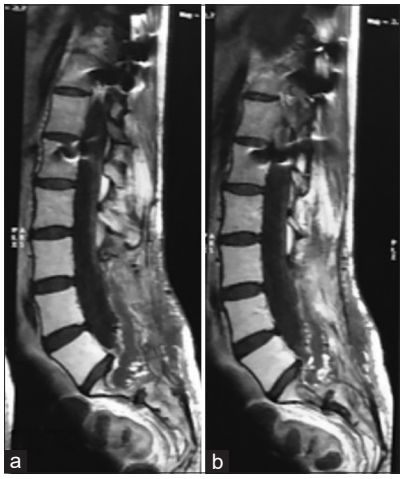

Figure 6: At 1-year follow-up, (a) T1-weighted sagittal MRI, small tumor recurrence in the sacrum.

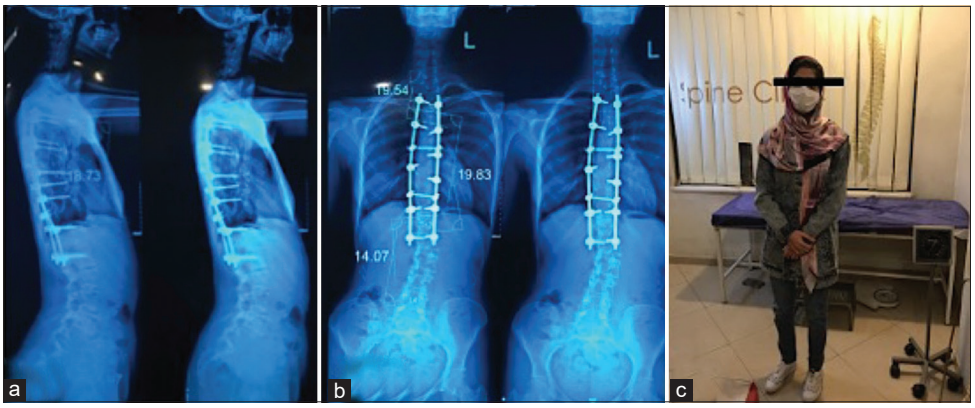

Figure 7: (a and b) Total spinal AP and lateral radiographs show persistent distal junctional disease (add-on) with no change in its degree. (c) Photograph of the patient with no apparent deformity.

immunohistochemical studies were confirmatory for an anaplastic MPE; the latter findings included; glial fibrillary acidic protein and S100 staining findings consistent with MPE [Figure 5]. Notably, the proliferative markers (i.e. iMIB-1 labeling index and Ki-67) emphasized that the tumor was highly malignant and would likely result in distant metastases.

Discharge and radiation therapy

The patient was discharged on the 6th postoperative day. She subsequently underwent radiotherapy. One year later, the patient had a follow-up sacral MRI scan that documented small residual/recurrent focal tumor [Figures 6 and 7]. She continued to exhibit residual incomplete saddle sensory deficit with urinary dysfunction requiring intermittent catheterization; otherwise she had no other new focal neurological deficits [Figure 6].

DISCUSSION

Intrasacral MPEs are quite rare.[1-3,4] They arise from the extradural remnants of the ependymal glia of the filum terminale, or they are a coccygeal medullary vestige.[1-3,4] These are slow growing and may become quite large before producing neurological symptoms/signs. Initially, these tumors fill and then gradually expand the sacral canal, ultimately extruding through the enlarged sacral foramina.[1-3,4] On very rare occasion, they may develop anaplastic features. Of the four similar cases found in the literature,[5-8] two were located over the sacrococcygeal bone subcutaneously and the other two were in the lumbar region compressing the cauda equina.[5-8]

Gross total excision is rarely feasible

Gross total removal (GTR) of these anaplastic lesions is almost impossible as they typically aggressively infiltrate the surrounding tissues. Therefore, radiation therapy is warranted even following a "presumed" GTR.[5-8] Note that for MPEs with anaplastic features, distant metastases are the most common clinical scenario, and these patients will likely follow a rapid course of neurological deterioration.

CONCLUSION

For patients presenting years after long scoliosis fusion surgery, complaints of pain should not be dismissed as simply "junctional" pathology. Here, a 23-year-old female presented 9 years following her scoliosis surgery with back pain and sphincter dysfunction newly attributed a sacral MPE with anaplastic features requiring aggressive surgical excision.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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