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Case Report

Extradural Myxopapillary Ependymoma in a 5 Year Old Child. A Case Report

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Abstract

Introduction: Myxopapillary ependymoma is a specific type of ependymoma that in very rare cases can occur in the extraspinal soft tissues. Categorized as grade I lesions myxopapillary ependymomas are considered benign but are capable of local recurrence, dissemination and a more aggressive disease course.

Case: The patient is a six year old boy with a formation in the sacral area with approximate dimensions 5 x 5 cm. Primary suspicion was hematoma because of history of repeated trauma to the buttocks. Ultrasonographic and computed tomography scans revealed the precise location, size and blood supply of the tumor. Gross- total resection was attempted.

Results: The sacrum and coccygeal bone were found to be intact and en-bloc resection was feasible. Histopathological result indicated extramedullary myxopapillary ependymoma. Postoperative period was uneventful. Follow-up magnetic resonance scan revealed normal postoperative finds and no signs of residual formation and/or recurrence.

Conclusion: Even though a very rare tumor, ME should be considered a possibility for sacral region tumors. Keeping in mind the potential of the tumor for local recurrence and metastases, it is advised that extradural subcutaneous ME be considered a low-grade malignant tumor and gross-total excision should be performed in all cases. Periodical postoperative examination is warranted for long term tumor control.

Keywords: *Extradural Ependymoma; Myxopapillary; Sacral Tumor*

Introduction

Myxopapillary ependymoma (ME) is a distinctive type of ependymoma that was first described as a separate pathological entity by Kernohan JW in 1932 [1]. It is a low-grade intradural tumor of ependymal origin that typically occurs in the conus medullaris and filum terminale of the spinal cord [2]. In the World Health Organization classification myxopapillary ependymomas are categorized as Grade I lesions [3,4]. Although ME are considered benign with a tendency for slow growth and local recurrence, especially after incomplete resection, they are capable of leptomeningeal dissemination and seldom even of extraneural metastases and a more aggressive disease course [5-7]. In some instances, ME may occur as a primary lesion in the extraspinal soft tissues [2]. Extradural ependymomas are very rare and are known to occur in several perisacral locations, as follows: 1) the extradural spinal canal with association with the dural part of filum terminale; 2) the bone substance of the sacrum; 3) the subcutaneous tissues dorsal to the sacrum [8-14]. Because of the erosive nature of the tumor it might be impossible to differentiate between one that starts within the sacrum and one that starts in the extradural spinal canal and erodes into the sacrum. Based on case reports in literature, the posterior subcutaneous location is the most common one for extradural ependymomas, followed by the presacral region [8,9,15,16].

Case Report

A six year old boy presented with a swelling in the sacral region with history of repeated blunt trauma to the area, as a result of falling on his buttocks. Initially hematoma was suspected because of the bluish coloration of the formation. The persisting lesion and its growth led the parents to seek medical attention. At the time of presentation the patient was complaint-free. Physical examination revealed a round shaped swelling in the sacral area soft tissues. Approximate dimensions 5 x 5 cm, relatively immobile in respect to adjacent tissues. Coloration- blue/red with tenderness on palpation (Figure 1). Ultrasonographic exam revealed a structure with high blood supply. Next step examination was contrast-enhanced Computed Tomography (CT) that presented a nodular lesion with irregular shape. Transverse dimensions 58mm, anterior-posterior: 29 mm; longitudinal: 55 mm. The tumor appeared to be very close to the coccygeal bone without signs of infiltrating it or the sacrum. Surrounding tissues also appeared intact. The density was homogenous with no significant change after intra venous contrast matter application- about 35 Hounsfield units. No necrotic areas were visualized. The vessel supplying the lesion was identified (Figure 2-4). Gross- total resection of the tumor was attempted. With the patient in prone position the approach was longitudinal incision along the lesion itself. The in-depth inspection revealed a tumor with lobular structure. The en-bloc resection was successful. The coccygeal bone was found to be intact. Express histopathological examination during operation indicated a benign formation (Figure 5 and 6). Postoperative period was uneventful. There was primary wound healing and the boy was discharged on the fifth postop day. Thorough histopathological examination revealed papillary, multinodular, hyper- cellular tumor constituted of atypical ependymal cells, arranged in perivascular pseudo- rosettes. Conclusion: extramedullary myxopapillary ependymoma, World Health Organization Grade I (Figure 7). Two weeks after the operation the patient underwent a Magnetic Resonance Tomographic scan. The results showed normal postoperative finds and no signs of residual formation and/or recurrence. The dural sac, conus medullaris and cauda equina were all intact (Figure 8 and 9).

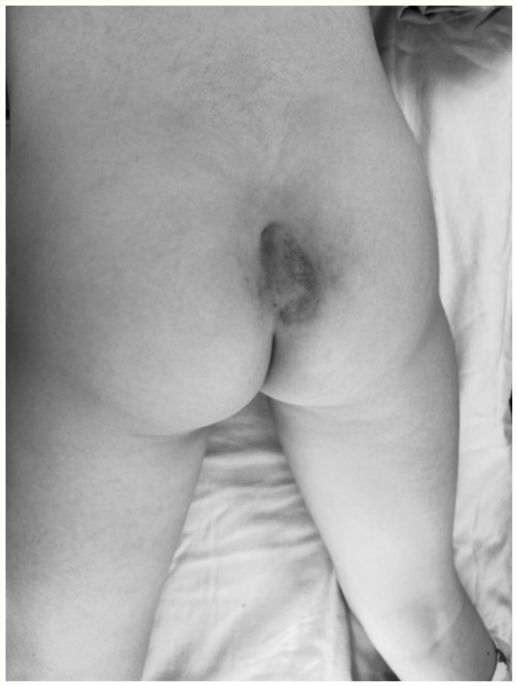

Figure 1: *The patient on the operating table in prone position with the formation clearly visible.*

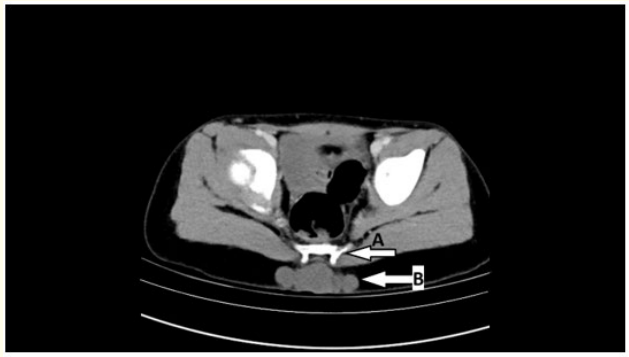

Figure 2: *Preoperative CT scan. A. Sacrum B. Tumor.*

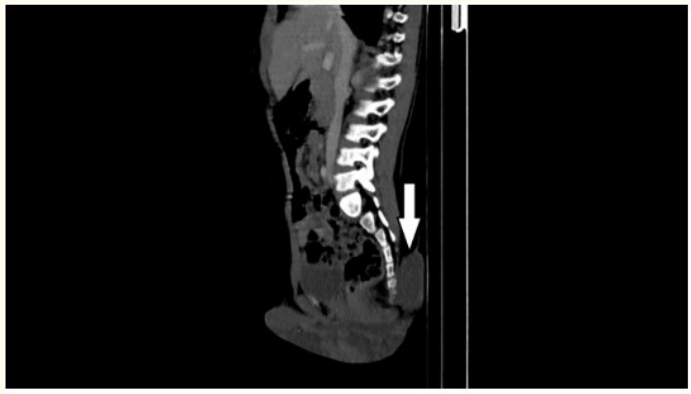

Figure 3: *Preoperative CT scan. Presents the tumor in sagittal view.*

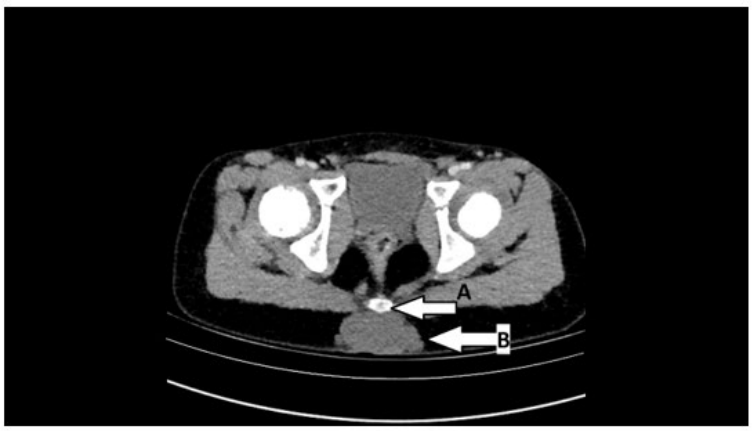

Figure 4: *Preoperative CT scan. A. Coccygeal bone B. Tumor.*

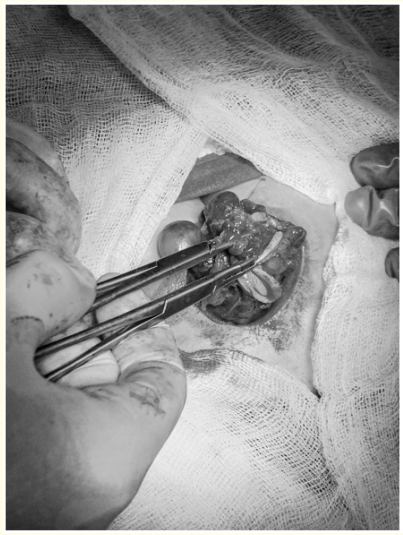

Figure 5: *Intraoperative view of the tumor.*

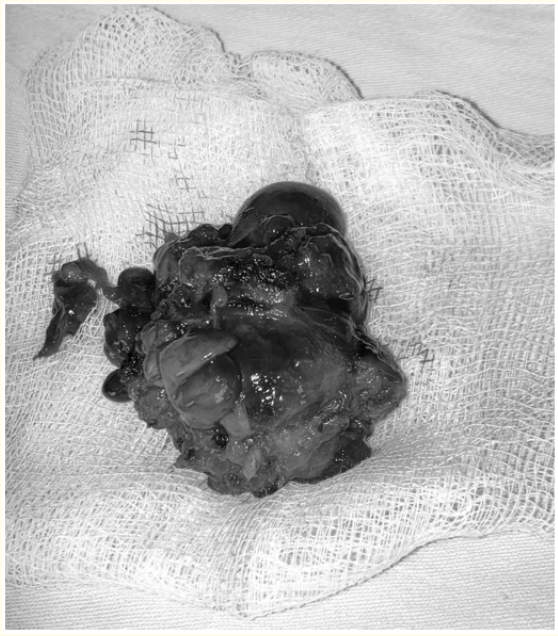

Figure 6: *The formation resected en-bloc.*

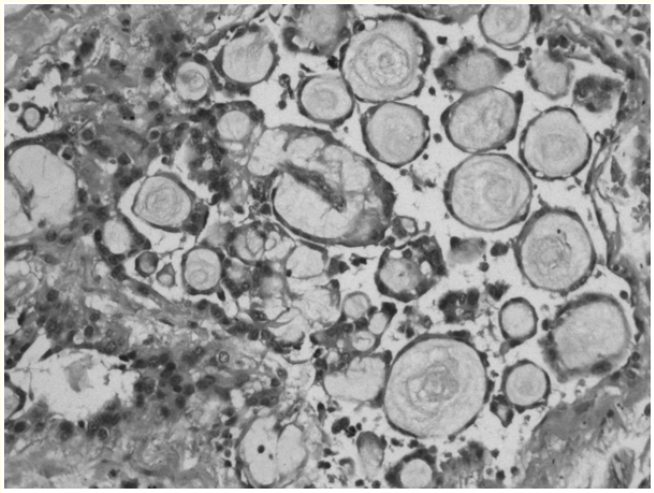

Figure 7: *Histopathological find.*

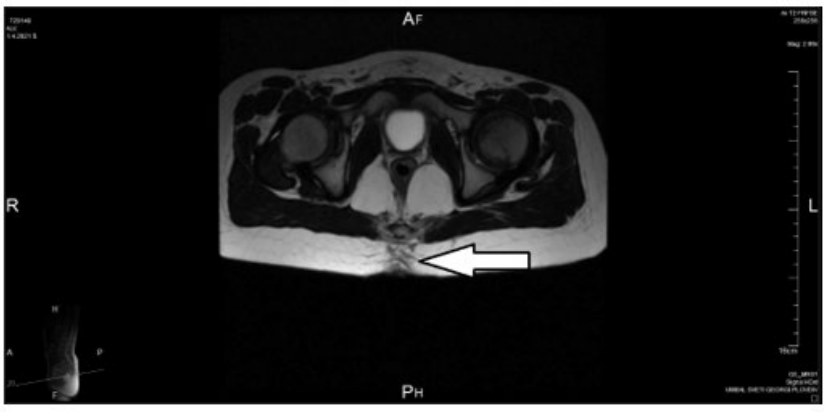

Figure 8: *Postoperative MRI. The arrow indicates the lesion site with no signs of recurrence or residual formation.*

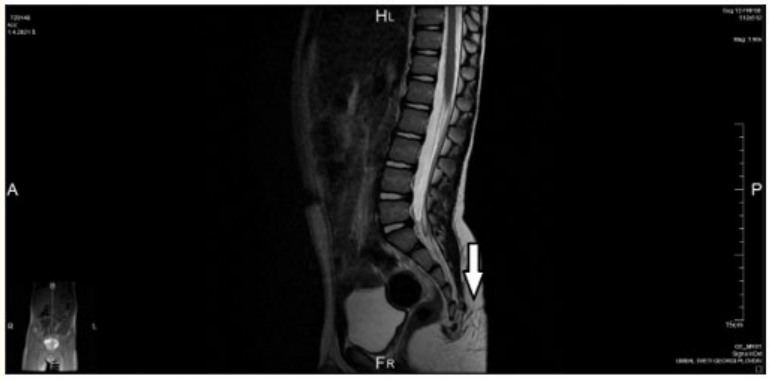

Figure 9: *Postoperative MRI. The arrow indicates the lesion site in sagittal view.*

Discussion

The surgical goal with all ependymomas, whether they are intra- or extradural, is gross-total resection when feasible [8,17]. Some authors have reported higher recurrence rates for tumors removed in a piecemeal fashion, even though a gross-total resection was achieved [18,19].

When the sacrum is involved, combined anterior-posterior approaches, either in the same sitting or during different operations, may be needed [9,13]. The role of radiation for lumbosacral ependymomas has not been adequately studied to warrant firm conclusions [8] and its efficacy in cases of gross-total resection and extradural ependymomas is controversial [1,5,9,15,17-23]. Extradural tumors are rare and so sufficient data is lacking on radiotherapy effectiveness. Most cases of local recurrence are treated with surgery and not radiation [8]. Metastatic disease to other organ systems typically does not respond to radiation therapy or chemotherapy [24]. A number of chemotherapeutic agents have been tried in in small series for recurrent spinal ependymomas, but no studies have shown compelling evidence to indicate that chemotherapy may be a primary treatment for these tumors [9,19,25,26]. Some case reports describe control of ME with chemotherapy [27,28], but sufficient data is lacking to draw any significant conclusions on this regimen [8]. Despite the significant risk of local recurrent and dissemination, the overall prognosis for lumbosacral intradural myxopapillary ependymomas appears to be very good. Authors report 94 - 95% 10- year survival rate [8,19,29]. Although rarely encountered in children, myxopapillary ependymomas have a more aggressive clinical course with increased risk of recurrence and dissemination when compared to the adult population. The reason for this more aggressive course in children is unclear [30].

Conclusion

Even though a very rare tumor, ME should be considered a possibility for sacral region tumors. Keeping in mind the potential of the tumor for local recurrence and metastases, it is advised that extradural subcutaneous ME be considered a low-grade malignant tumor and timely gross-total excision should be performed in all cases. Periodical postoperative examination is warranted for long term tumor control.

Bibliography

1. Kernohan JW. "Primary tumors of the spinal cord and intradural filum terminale". *Cytology and Cellular Athology of the Nervous System* 3 (1932): 993-1025.

2. Zaidi SN., *et al*. "Myxopapillary ependymoma masquerading as subcutaneous saccroccygeal non-healing ulcer: Case report". *Annals of Saudi Medicine* 34.3 (2014): 262-264.

3. Shaw EG., *et al*. "Radiotherapeutic management of adult intraspinal ependymomas". *International Journal of Radiation Oncology, Biology, Physics* 12.3 (1986): 323-327.

4. Wiestler OD., *et al*. "Ependymomas". In Kleihues P, Cavernee WK (eds): Pathology and Genetics of Tumors of the Nervous System, ed 2. Lyon: IARC Press (2000): 50-56.

5. Chinn DM., *et al*. "Management of children with metastatic spinal myxopapillary ependymoma using craniospinal irradiation". *Medical and Pediatric Oncology* 35.4 (2000): 443-445.

6. Gagliardi FM., *et al*. "Ependymomas of the filum terminale in childhood: report of four cases and review of the literature". *Child's Nervous System* 9.1 (1993): 3-6.

7. Nagib MG and O'Fallon MT. "Myxopapillary ependymoma of the conus medullaris and filum terminale in the pediatric age group". *Pediatric Neurosurgery* 26.1 (1997): 2-7.

8. Fassett DR and Schmidt MH. "Lumbosacral ependymomas: a review of the management of intradural and extradural tumors". *Neurosurgical Focus* 15.5 (2003): E13.

9. Fourney DR., *et al*. "Intraspinal extradural myxopapillary ependymoma of the sacrum arising from the filum terminale externa. Case report". *Journal of Neurosurgery* 93.2 (2000): 322-326.

10. Morantz RA., *et al*. "Extraspinal ependymomas. Report of three cases". *Journal of Neurosurgery* 51.3 (1979): 383-391.

11. Helwig EB and Stern JB. "Subcutaneous sacrococcygeal myxopapillary ependymoma. A clinicopathologic study of 32 cases". *American Journal of Clinical Pathology* 81.2 (1984): 156-161.

12. Lynch J., *et al*. "A sacrococcygeal extraspinal ependymoma in a 67-year-old man: a case report and review of the literature". *British Journal of Plastic Surgery* 55.1 (2002): 80-82.

13. Morantz RA. "Ectopic ependymoma of the sacrococcygeal region". In Doty JR, Rengachary SS (eds): Surgical Disorders of the Sacrum. New York: Theime Medical (1992): 177-179.

14. Schurmann K and Wallenfang T. "Rare sacral space-occupying lesions, their surgical management and reconstructive measures involved". *Acta Neurochirurgica* 92.1-4 (1988): 106-117.

15. Gerston KF., *et al*. "Presacral myxopapillary ependymoma presenting as an abdominal mass in a child". *Journal of Pediatric Surgery* 20.3 (1985): 276-278.

16. Pulitzer DR., *et al*. "Subcutaneous sacrococcygeal ("myxopapillary") ependymal rests". *American Journal of Surgical Pathology* 12.9 (1988): 672-677.

17. Lemberger A., *et al*. "Sacrococcygeal extradural ependymoma". *Cancer* 64.5 (1989): 1156-1159.

18. Waldron JN., *et al*. "Spinal cord ependymomas: a retrospective analysis of 59 cases". *International Journal of Radiation Oncology, Biology, Physics* 27.2 (1993): 223-229.

19. Sonneland PR., *et al*. "Myxopapillary ependymoma. A clinicopathologic and immunocytochemical study of 77 cases". *Cancer* 56.4 (1985): 883-893.

20. Celli P., *et al*. "Ependymoma of the filum terminale: treatment and prognostic factors in a series of 28 cases". *Acta Neurochirurgica* 124.2-4 (1993): 99-103.

21. Ross DA., *et al*. "Myxopapillary ependymoma. Results of nucleolar organizing region staining". *Cancer* 71.10 (1993): 3114-3118.

22. Schild SE., *et al*. "The results of radiotherapy for ependymomas: the Mayo Clinic experience". *International Journal of Radiation Oncology, Biology, Physics* 42.5 (1998): 953-958.

23. Schweitzer JS and Batzdorf U. "Ependymoma of the cauda equina region: diagnosis, treatment, and outcome in 15 patients". *Neurosurgery* 30.2 (1992): 202-207.

24. Kramer GW., *et al*. "Subcutaneous sacrococcygeal ependymoma with inguinal lymph node metastasis". *Journal of Neurosurgery* 68.3 (1988): 474-477.

25. Chamberlain MC. "Salvage chemotherapy for recurrent spinal cord ependymoma". *Cancer* 95.5 (2002): 997-1002.

26. Timmerman W and Bubrick MP. "Presacral and postsacral extraspinal ependymoma. Report of a case and review of the literature". *Diseases of the Colon and Rectum* 27.2 (1984): 114-119.

27. Madden JR., *et al*. "Experience with tamoxifen/etoposide in the treatment of a child with myxopapillary ependymoma". *Medical and Pediatric Oncology* 37.1 (2001): 67-69.

28. Fujiwara Y., *et al*. "Remarkable efficacy of temozolomide for relapsed spinal myxopapillary ependymoma with multiple recurrence and cerebrospinal dissemination: a case report and literature review". *European Spine Journal* 27.3 (2018): 421-425.

29. Mork SJ and Loken AC. "Ependymoma: a follow-up study of 101 cases". *Cancer* 40.2 (1977): 907-915.

30. Bagley CA., *et al*. "Long term outcomes following surgical resection of myxopapillary ependymomas". *Neurosurgical Review* 32.3 (2009): 321-334.

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