Subcutaneous Intergluteal Myxopapillary Ependymoma in a Child

Bir Çocukta Subkutan İntergluteal Miksopapiller Ependimom

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ABSTRACT

Myxopapillary ependymomas are mostly seen in adult population and derived from ependymal remnants, but it is rarely seen in childhood. The mass was totally excised from sacrococcygeal area. The pathological diagnosis was reported as subcutaneous myxopapillary ependymoma (WHO grade 1). Myxopapillary ependymomas can be excised safely with minimal morbidity.

Key Words: Child, Ependymoma, Myxopapillary

ÖZ

Miksopapiller ependimomlar; ependimal kalıntılardan oluşur, çoğunlukla erişkin popülâsyonda görülür ancak çocukluk çağında da nadir görülür. Bu yazıda, sakrokoksik alanda ağrılı semi-solid kitlesi olan ve patolojik raporu olan 13 yaşında bir erkek çocuk sunulmaktadır. Kitle tamamen sakrokoksigeal bölgeden çıkarıldı. Patolojik tanısı subkutan miksopapiller ependimom olarak rapor edildi. (WHO grade 1). Miksopapiller ependimomlar minimal morbidite ile güvenle eksize edilebilir.

Anahtar Kelimeler: Çocuk, Ependimom, Miksopapiller

INTRODUCTION

Myxopapillary ependymomas are mostly seen in adult population and derived from ependymal remnants, but it is rarely seen in childhood. The presence of anaplasia is not known. Myxopapillary ependymomas consist of 13% of all spinal ependymomas. In this case report, we aimed to report case of myxopapillary ependymoma in a child.

CASE REPORT

A 13 years old boy patient applied to our outpatient clinic with a pain and swelling in sacrococcygeal area. He had these complaints about 18 months. In his physical examination a firm, well circumscribed immobile lesion (mass?) was palpated on midline in sacrococcygeal area (Figure 1). There was no neurological deficit. A semi solid, 5x1.5x4 cm in size, densely content lesion was seen in ultrasound imaging. Epidermoid cyst was thought as early diagnosis and after that 6x3x3.5 in size mass was totally excised from sacrococcygeal area (Figure 2). The pathological diagnosis was compatible with subcutaneous myxopapillary ependymoma (WHO grade 1) and the tissue was GFAP, vimentin and s100 positive in immunohistochemical examination.The patient was also followed up by oncology department and his abdominal ultrasound imagings, which were performed 10 days and 2 months after excision, were normal. A lumbosacral spinal MRI was performed after 4 months and there was not any sign of pathological contrast uptake.

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Ethics Committee Approva / *Etik kurul onayı:* *During the registration, an informed consent was signed by the parents for participation in the research.* ***Onay:*** Kayıt sırasında veliler tarafından araştırmaya katılım için bilgilendirilmiş bir onay imzalanmıştır.

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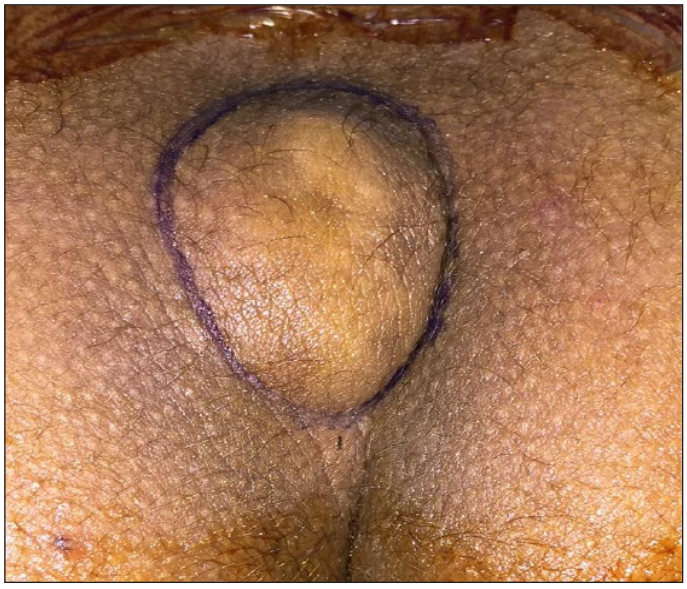

Figure 1: Figure 1: The photography of immobile palpable mass in sacrococcygeal area.

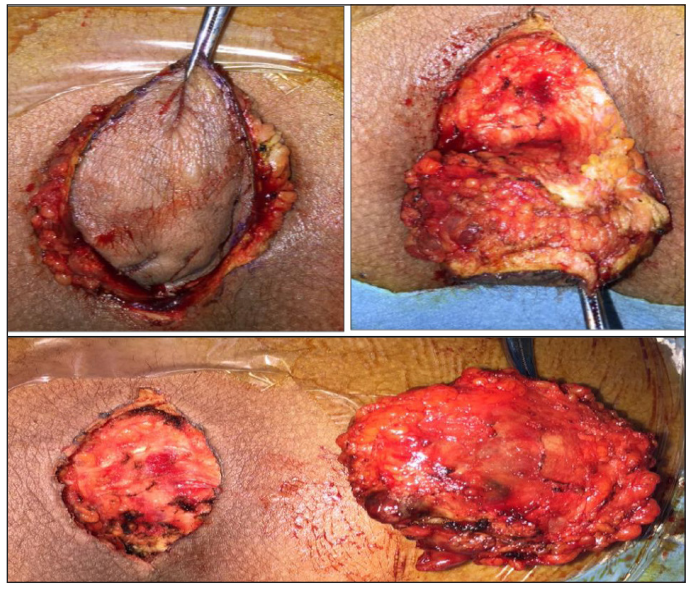

Figure 2: The photos of surgery.

DISCUSSION

Extraspinal sacrococcygeal subcutaneous ependymoma was first reported by Mallory in 1902 (1). Although in the literature the age range varies between 2 months and 67 years, the mixopapillary ependymomas are seen rare in childhood (1–9). Ependymal tumors are divided into 4 groups according to WHO classification as the following; 1. Subependymoma, 2. Myxopapillary ependymoma, 3. Ependymoma and 4. Anaplastic ependymoma. Myxopapillary ependymoma is the most commonly seen group and they are classified as grade 1 tumor according to WHO and are located near the gluteal cleft, which leads to a common clinical misdiagnosis of pilonidal disease (4, 6,7,10). The exact origin of this tumor remains unknown. Most believe that, this is a tumor arising from the coccygeal medullary vestige, but the other hypothesize is that this tumor arises from a persistence of the neuroenteric canal or from heterotopic ependymal cell rests located within the region (4). Their prognosis is benign and growth rate is slow. The clinical presentation depends on the location of the tumor, with those located posterior to the sacrum most commonly presenting with pain. Our case complained of pain and swelling in sacrococcygeal area in consistent with literature. Diagnostic imaging generally consists of standard radiographs, ultrasonography and Magnetic Resonance Imaging (MRI) examination of the sacrum, pelvis and lumbosacral spine. In our case ultrasonography was used. The differential diagnosis for posteriorly located lesions include pilonidal cyst, lipoma, chordoma, teratoma, dermoid cysts, or neurofibroma. Inflammatory lesions such as perirectal abscess and internal fistula may also be seen (4). Recommended treatment of subcutaneous sacrococcygeal ependymoma is wide local excision with negative margins (3). The mass of our case was excised totally in consistent of literature. Current treatment of newly diagnosed ependymomas in children that are completely resected do not include chemotherapy (9). In cases of margin-positive disease, adjuvant radiation therapy has proven useful in several case studies (2,3). Pathologic examination typically demonstrates positive immunohistochemistry for GFAP and S-100 (5,6). The pathology materials of tissue was GFAP, vimentin and s100 positive in immunohistochemical examination. Subcutaneous sacrococcygeal myxopapillary ependymomas have high recurrance rate but they rarely metastate to distant tissues, so prolonged surveillance is required (2,6,7). Survival rate after total or partial resection is more than 10 years. In the followup our case, we have not see any recurrences and complication. In conclusion; myxopapillary ependymoma is rarely seen in childhood and it has to be thought in differential diagnosis with sacrococcygeal area pathologies. It can be excised safely with minimal morbidity.

REFERENCES

1. Mallory FB. Three gliomata of ependymal origin: two in the fourth ventricle, one subcutaneous over the coccyx. J Med Res 1902; 8:1–10.
2. Shuangshoti S, Rushing EJ, Mena H, Olsen C, Sandberg GD. Supratentorial extraventricular ependymal neoplasms. Cancer 2005;103:2598–605.
3. Chou S, Soucy P, Carpenter B. Extraspinal ependymoma. J Pediatr Surg 1987;22: 802-3.
4. Morantz RA, Kepes JJ, Batnitzky S, Masterson BJ. Extraspinal ependymoma: report of 3 cases. J Neurosurg 1979;51: 383-91.
5. Johnson JM, Jessurun J, Leonard A. Sacrococcygeal ependymoma: case report and review of the literature. J Pediatr Surg 1999; 34: 1405-7.

6. Ma YT, Ramachandra P, Spooner D. Primary subcutaneous sacrococcygeal ependymoma: a case report and review of the literature. Br J Radiol 2006; 79: 445-7.
7. Lynch J, Kelly N, Fitzpatrick B, Regan P. A sacrococcygeal extraspinal ependymoma in a 67-year-old man: a case report and review of the literature. J Plast Reconstr Aesthet Surg 2002; 55: 80-2.
8. Timmerman W, Bubrick MP. Presacral and postsacral extra-spinal ependymoma: report of a case and review of the literature. Dis Colon Rectum 1984;27:114-9.
9. Amin R, Berdan E, Knipstein J Jarzembowski J, Siddiqui S. Extraspinal sacrococcygeal ependymoma masquerading as sacrococcygeal teratoma in the pediatric patient. Pediatr Surg Int 2018;34:109-12.
10. Mc Eachron K, Gaertner W. Extradural sacrococcygeal subcutaneous ependymoma misdiagnosed as pilonidal disease: case report and review of the literature. J Surg Case Rep 2016;7:1-3.