Asian Journal of Orthopaedic Research

Volume 6, Issue 2, Page 58-63, 2023; Article no.AJORR.101988

Extraspinal Sacrococcygeal Ependymoma

Akcali Ayhan [a++], Ozyigit Elif [b#], Ozyigit Omer [c++] and Ekinci Safak [a†*]

[a] Private A Hospital, Kocasinan, Bahçelievler, Istanbul, Turkey.
[b] Kanuni Sultan Süleyman Training and Research Hospital, Istanbul, Turkey.
[c] Atlas University Medicine Hospital, Güneşli, Istanbul, Turkey.

Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

Article Information

Open Peer Review History:
This journal follows the Advanced Open Peer Review policy. Identity of the Reviewers, Editor(s) and additional Reviewers, peer review comments, different versions of the manuscript, comments of the editors, etc are available here: https://www.sdiarticle5.com/review-history/101988

Case Report

Received: 28/04/2023
Accepted: 09/07/2023
Published: 21/07/2023

ABSTRACT

Ependymomas usually arise within the spinal cord or in the brain. Ependymomas generally originated in the sacrum or extraspinal parasacral soft tissues and they are rare tumors. We reported a case report in the subcutaneous tissue posterior to the sacrococcygeal region. Radiologic and pathologic studies are presented. Differantial diagnosis and the pathogenesis of extraspinal ependymomas are discussed.

Keywords: Ependymoma; lomber spine; sacrococcygeal region.

1. INTRODUCTION

Ependymal tumors usually arise within the spinal cord or in the brain. Spinal ependymomas are 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.

[++] M.D. Radiology;
[#] M.D. Physical Therapy and Rehabilitation;
[†] M.D. Associate Professor Orthopedics and Traumatology;
**Corresponding author: Email: safakekinci@yahoo.com;*

The cellular ependymoma is predominately an infratentorial tumor that occurs mainly in children and adolescents. It typically fills the forth 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 ependimomas 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 lesions at the caudal forth 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].

2. 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, addmitted to our hospital because of the presence of a slow growing mass in the intergluteal fold for two years. On physical examination demonstrated 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 (Fig. 2).

A magnetic resonence 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 subcutenous soft tissue, unattached to the sacrum and with no connection the spinal canal. Complete local excision was performed.

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

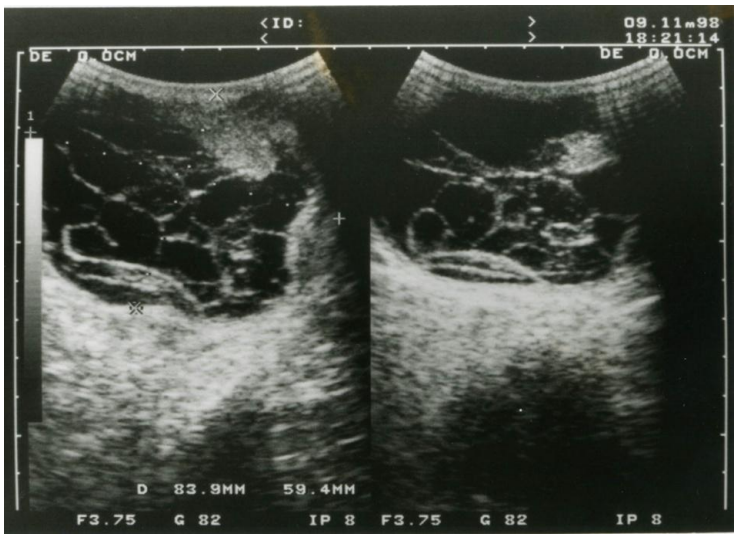

Fig. 1. Ultrasonography shows well defined cystic lesion (85x60 mm in diameter) which contain multiseptal and with solid component next to the wall

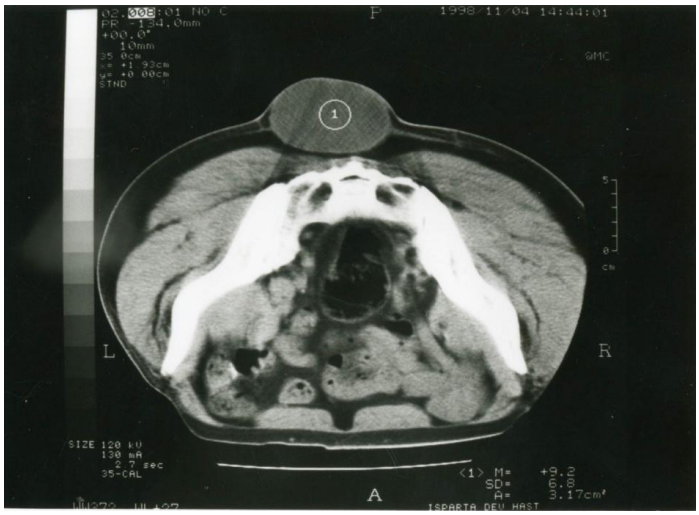

Fig. 2. CTscan of the pelvis shows 85x60 mm in diameter, at 9 HU density hypodens mass posterior of the sacrum, in subcutaneous fatty tissue

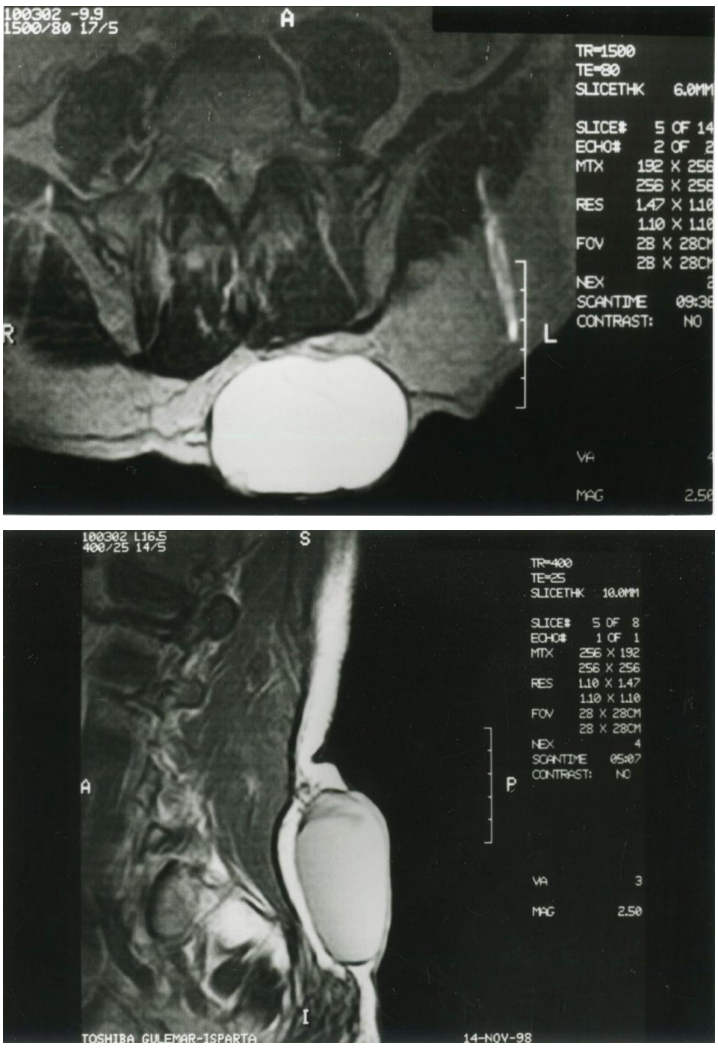

Fig. 3a-b. MR images show the cystic nature of the lesion, with hyperintensity on T2/Weighted images and hypointensity on T1/Weighted images and nonhomogenous intensity at superior of the cystic lesion

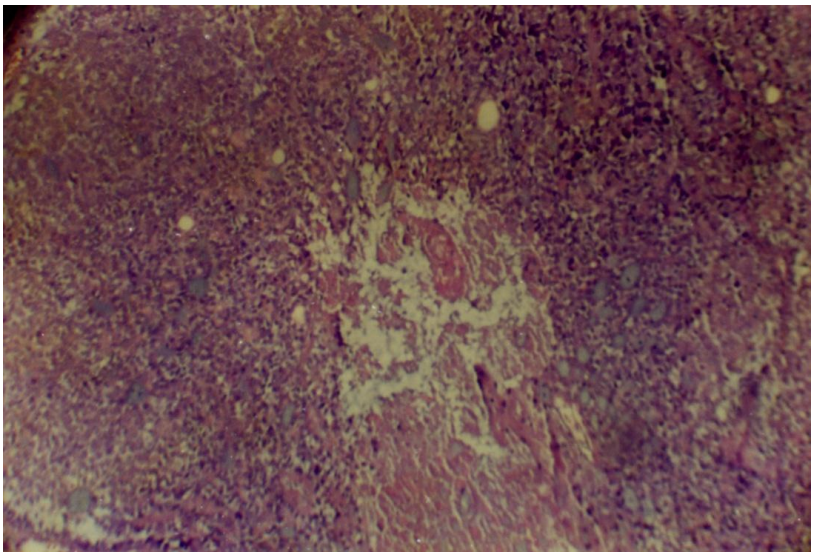

Fig. 4. The true ependymal rosettes contain a well defined central lumen, necrosis and blue-tinged musin microcysts

Immunopeoxidas studies on formalin –fixed, paraffin embeddet tumor tissue showed positive staining for wimentin and glial fibrillary asidic protein and lack of staining for cytokeratins, S-100 protein and neuron specific enalase. The lightmisroscopic and immunohistochemical data were considered diagnostic of myxopapillary epandymoma.

3. DISCUSSION

"Ependymomas are rare neuroepithelial neoplasms accounting for 1.9% of primary CNS tumors and up to 60% of primary tumors of the spinal cord, in adults. MPEs represent almost 27% of all ependymomas and are primarily intradural extramedullary tumors located more often in the lumbar thecal sac in proximity to the conus medullaris, cauda equina or filum terminale (intraspinal variant)" [6].

"A few common localizations are the pre-or post-sacral regions, in the so-called extraspinal variant. They generally arise in the 4thdecade of life and only 8–20% occur in children" [7,8].

"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" [9,10].

"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" [11].

"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" [12,13,14]. "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" [15,16]. "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" [17,18,19,20].

"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" [21].

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 [22].

4. CONCLUSION

Spinal ependymomas may rarely arise from heterotopic ependymal cell clusters and thus occur in an extraspinal location. The presentation of our case and a review of the literature demonstrate that these tumors have distinct radiographic and clinical characteristics. They occur mainly in patients in the third-fourth decade of life, and present either in the soft tissue posterior to the sacrum or in the pelvis. In the case of posterior tumors, the patient exhibits a mass which is usually mistaken for a pilonidal cyst, sacrococcoccygeal teratom and neurogenic tumor. Ultrasonography, computerized tomography studies and MRI should be used differential diagnosis. The preferred method is to completely remove the tumor. Radiation therapy should be used if this is not feasible. Because of the increased incidence of systemic metastases, this tumors must be follow up the average postoperative period approximately 10 years.

CONSENT

As per international standard or university standard, patient(s) written consent has been collected and preserved by the author(s).

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author(s).

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Schweitzer JS, Batzdorf U. Ependymoma of the cauda equina region; diagnosis, treatment and outcome in 15 patients. Neurosurgery. 1992;30:202-207.
2. Osborn AG. Diagnostic Neuroradiology in brain tumors and tumorlike masses. Mosby year Book, Inc. Washington D.C. 1994; 906-909.
3. Miralbell R, Louis DN, O'Keeffe D, Rosenberg AE, Suit HD. Metastatic ependymoma of the sacrum. Cancer. 1990;65:2353-2355.
4. Inceoglu R, Ozer F, Pamir N, Küllü S. Extraspinal ependymoma presenting as a subcuteneus mass posterior to the sacrococcygeal region. Case Report. Paraplegia. 1993;31:800-802.
5. Rengachary. Masses of the sacrum. In; Wilkins HR, Rengachary SS (Editors) Neurosurgery. Volume III McGraw-Hill Inc. New York. 1985;1079-1083.
6. Louis DN, Perry A, Wesseling P, Brat DJ, Cree IA, Figarella-Branger D, Hawkins C, Ng HK, Pfister SM, Reifenberger G, et al. The 2021 WHO classification of tumors of the central nervous system: A summary. Neuro-Oncology. 2021;23:1231–1251.
7. Hamilton KR, Lee SS, Urquhart JC, Jonker BP. A systematic review of outcome in intramedullary ependymoma and astrocytoma. J. Clin. Neurosci. 2019;63: 168–175.
8. Marchesini N, Tommasi N, Faccioli F, Pinna G, Sala F. Cauda equina ependymomas: Surgical treatment and long-term outcomes in a series of 125 patients. J. Neurosurg. Spine. 2022;36: 452–463.
9. Ma YT, Ramachandra P, Spooner D. Case report: Primary subcutaneous sacrococcygeal ependymoma: A case report and review of the literature. Br J Radiol. 2006;79(941):445-447.
10. Lee KJ, Min BW, Seo HJ, Cho CH. Subcutaneous sacrococcygeal myxopapillary ependymoma in Asian female: A case report. J Clin Med Res. 2012;4(1):61-63.
11. Keslar PJ, Buck JL, Suarez ES. Germ cell tumors of the sacrococcygeal region: Radiologic-pathologic correlation. Radiographics. 1994;14:607–620.
12. Yust Katz S, Cachia D, Kamiya-Matsuoka C, Olar A, Theeler B, Penas Prado M, Gilbert MR, Armstrong T. Ependymomas arising outside of the central nervous system: A case series and literature review. J. Clin. Neurosci. 2018;47:202–207.
13. Schiavello E, Biassoni V, Antonelli M, Modena P, Cesaro S, Pierani P, Gandola

L. Pediatric Extraspinal Sacrococcygeal Ependymoma (ESE): An Italian AIEOP experience of six cases and literature review. Childs Nerv. Syst. 2018;34:1291–1298.
14. Batich KA, Riedel RF, Kirkpatrick JP, Tong BC, Eward WC, Tan CL, Pittman PD, McLendon RE, Peters KB. Recurrent extradural myxopapillary ependymoma with oligometastatic spread. Front. Oncol. 2019;9:1322.
15. Bale PM. Ependymal rests and subcutaneous sacrococcygeal ependymoma. Pathology. 1980;12:237–243.
16. Maiorana A, Fante R, Fano RA. Myxopapillary ependymoma of the sacrococcygeal region. Report of a Case. Pathologica. 1989;81:471–476.
17. Lemberger A, Stein M, Doron J, Fried G, Goldsher D, Feinsod M. Sacrococcygeal extradural ependymoma. Cancer. 1989;64: 1156–1159.
18. Bell DA, Woodruff JM, Scully RE. Ependymoma of the broad ligament. A report of two cases. Am. J. Surg. Pathol. 1984;8:203–209.
19. Duggan MA, Hugh J, Nation JG, Robertson DI, Stuart GC. Ependymoma of the uterosacral ligament. Cancer. 1989;64: 2565–2571.
20. Kleinman GM, Young RH, Scully RE. Ependymoma of the ovary: Report of three cases. Hum. Pathol. 1984;15:632–638.
21. Alexander Muacevic, John R. Adler, Subramaniam Ramkumar, Cliff A. Wanniang, Anju Risa Wahlang, Alepes Lamin JC. Subcutaneous sacro coccygeal myxopapillary ependymoma: A case report and a comprehensive review of the literature reappraising its current diagnostic approach and management. Cureus Journal of Medical Science. 2021 May; 13(5):e14931.
22. Kyung-Jae Leea, Byung-Woo Mina, Hyuk-Jun Seoa, Chul-Hyun Choa. Subcutaneous sacrococcygeal myxopapillary ependymoma in Asian female: A case report. J Clin Med Res. 2011;4(1):61-63.

Peer-review history:
The peer review history for this paper can be accessed here:
https://www.sdiarticle5.com/review-history/101988