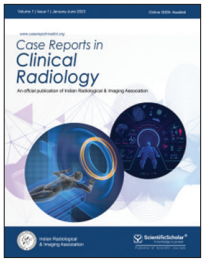
ScientificScholar®
Knowledge is power
Publisher of Scientific Journals

www.caserepclinradiol.org

Case Reports in Clinical Radiology



Case Report

Imaging in sacrococcygeal subcutaneous myxopapillary ependymoma: Sporadically reported from Asian population

Chetankumar Vajerambhai Dhandhalya[1], Shilpa L. Chudasama[1], Hiral Kishorbhai Pansuriya[1]

[1]Department of Radiodiagnosis, Shri M P Shah Government Medical College, G G Government Hospital Campus, Jamnagar, Gujarat, India.

***Corresponding author:**
Hiral Kishorbhai Pansuriya, Department of Radiodiagnosis, Shri M. P. Shah Government Medical College, G. G. Government Hospital Campus, Jamnagar, Gujarat, India.

hiripansuriya@gmail.com

Received: 18 April 2023
Accepted: 10 May 2023
EPub Ahead of Print: 03 June 2023
Published: 13 July 2024

DOI
10.25259/CRCR_74_2023

Quick Response Code:

ABSTRACT

Subcutaneous myxopapillary ependymoma in sacrococcygeal region is an unusual neoplasm. It usually affects younger age groups. Only sporadic case reports from the Asian population have been reported. We here report a case of subcutaneous sacrococcygeal myxopapillary ependymoma suspected at ultrasound, suggested at magnetic resonance imaging examination, and confirmed histologically in a 10-year-old boy. We aimed at explaining its pathology, clinical implications, management, and relevant literature review.

Keywords: Sacrococcygeal ependymoma, Rare extraspinal subcutaneous ependymoma, Myxopapillary ependymoma

INTRODUCTION

Ependymomas outside the spine are uncommon, with most of them being subcutaneous in the sacrococcygeal region. It is uncommonly reported from Asian population, however no race variation is noted.

CASE REPORT

A 11-year-old boy presented at surgical outpatient department with complain of natal cleft swelling with discomfort. Clinical examination revealed a non-tender mobile subcutaneous swelling of approximate size (4 × 8) cm with no apparent abnormality of the overlying skin [Figure 1]. As part of further evaluation local part, ultrasound examination was performed. A well-defined heterogeneously hypoechoic lesion with significant internal vascularity was found [Figure 2]. Pelvic and lumbo-sacro-coccygeal magnetic resonance imaging (MRI) examination was done for lesion characterization and delineation of its extent. MRI examination demonstrated a well-defined, well-circumscribed subcutaneous intermediate T2 and high short tau inversion recovery signal intensity lesion of size (4.8 × 2.9 × 6.5) cm (AP × TRA × CC) with multiple internal hypointense septae, and well-defined hypointense septae capsule in sacrococcygeal region at upper edge of intergluteal cleft. Lesion showed minimal diffusion restriction on diffusion-weighted imaging (with minimal low signal on corresponding apparent diffusion coefficient maps). Intense enhancement on post-contrast T1-weighted MRI with non-enhancing internal fibrous septae and peripheral fibrous capsule was evident [Figure 3]. The lower dural sac, cauda equina, and cone were found to be unremarkable. Radiologically, subcutaneous myxopapillary ependymoma was diagnosed based on MRI features and was confirmed histologically [Figure 4].

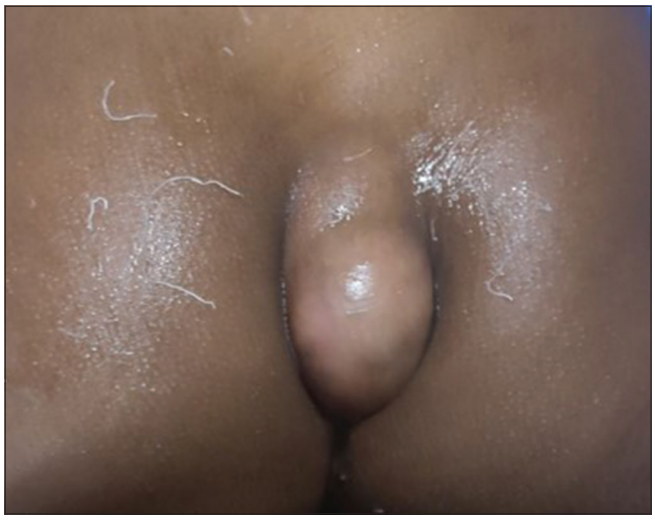


Figure 1: Clinical examination image showing swelling in intergluteal cleft region.

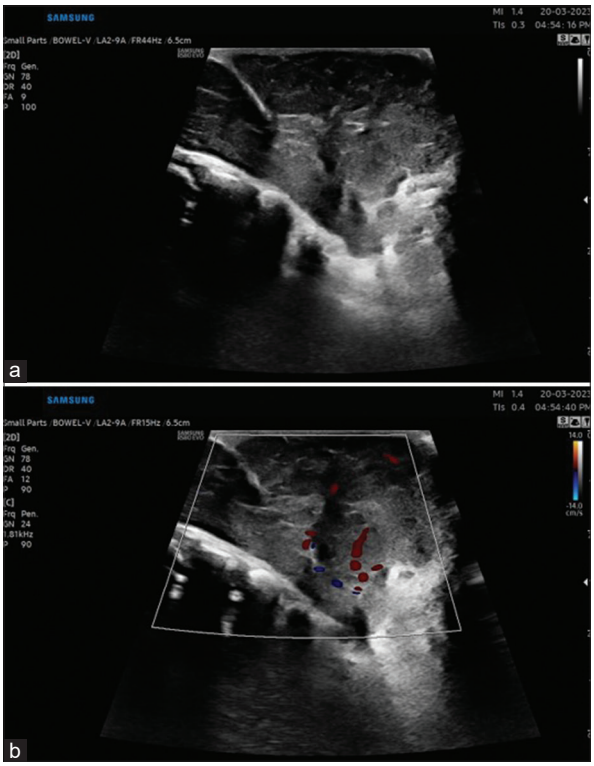


Figure 2: (a) Sagittal gray scale ultrasound image showing well-defined heterogeneously hypoechoic lesion subcutaneously in sacrococcygeal region at upper edge of intergluteal cleft (b) lesion demonstrated internal vascularity on color Doppler ultrasound.

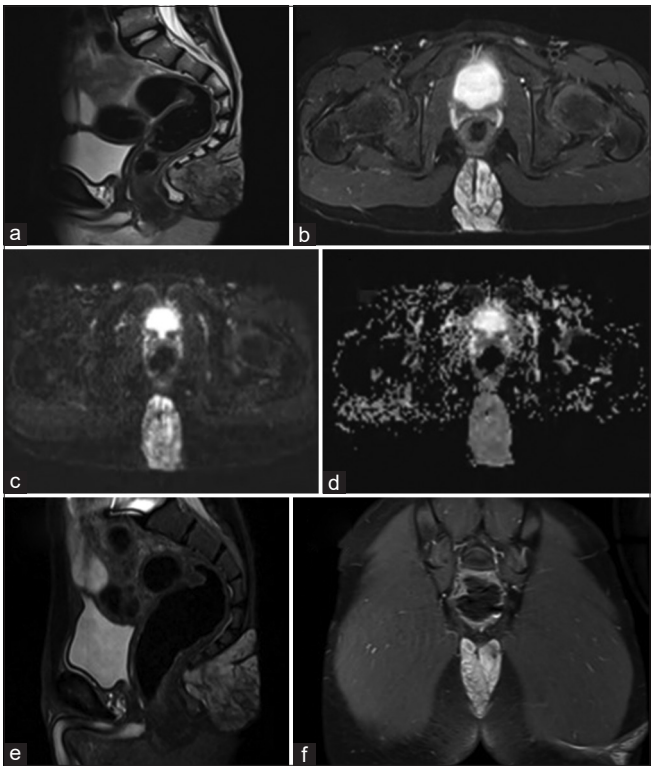


Figure 3: Magnetic resonance images (a-f): Lesion in subcutaneous plane in sacrococcygeal region at upper edge of intergluteal cleft shows intermediate signal on (a) T2W sagittal image, (b) high signal on short tau inversion recovery axial image, with hypointense capsule and septa. Minimal diffusion restriction with low signal on corresponding apparent diffusion coefficient maps (c and d) and intense enhancement with non-enhancing capsule and septa on T1W post-contrast images (e) sagittal and (f) coronal.

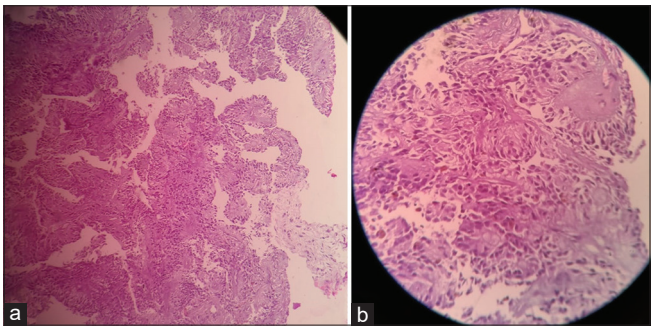


Figure 4: Histology images (a and b) H and E-stained sections show tumor tissue arranged in papillary pattern, Papillae show central fibrovascular core, at places stroma show bluish myxoid like areas. Papillae are lined by cells having round nuclei with elongated cytoplasm. No evidence of mitosis, necrosis or microvascular proliferation is seen. Overall findings suggest histology of myxopapillary ependymoma. H and E: Hematoxylin and eosin.

DISCUSSION

The rare occurrence of ependymoma outside the spine is probably due to heterotopic sites having the ependymal cells. Four general scenarios exist for the ependymomas outside the spine. (i) As metastasis from a primary central nervous system (CNS) ependymoma, (ii) primary spinal cord,

cauda equina, or filum terminale ependymoma extending directly into the sacrococcygeal soft tissues, (iii) primary cutaneous and subcutaneous neoplasm without spinal cord communication, and (iv) primary tumor of abdominal, pelvic, or presacral region.[1,2] The third scenario holds true in our case. Myxopapillary ependymomas are now classified as the World Health Organization Grade II CNS neoplasms instead of Grade I because the prognosis is now similar to traditional ependymomas.[3] The myxopapillary variant of ependymoma is an extremely rare type of ependymoma. These well-circumscribed tumors with slow growth rate can occur at any age mainly in young adults with the mean age of diagnosis being 36 years.[4] Ependymomas outside the spine in the sacrum are presacral or retro sacral masses. The most likely extraneural pathways for ependymal tumors are likely to be the lymphatic or hematogenous spread.[5] A presacral mass can cause mass effect on the bladder or bowel, and a post-sacral mass can present as swelling of the buttocks.[6] The diagnostic technique of choice for the diagnosis, grading, and staging of these tumors is MRI. The management of the ependymoma is determined according to the grading and the tumor site. Metastases were more frequent in very rare extrathecal post sacral (18%) than in presacral ependymomas (7%). Pulmonary, skeletal, and regional nodal and hepatic metastasis occurs in sacrococcygeal myxomapillary ependymoma.[5] Complete surgical removal is the key to curative treatment. Metastatic disease, incomplete removal, and coccygeal involvement require coccygectomy and post-surgical radiotherapy.[7,8] Due to risk of metastasis and local recurrence, long duration close follow-up is required. Complete removal at the initial operation provides the best survival outcome in patients with these tumors.[9] We report a case of a subcutaneous sacrococcygeal myxopapillary ependymoma in a young boy. As far as we know, rare previous sporadic reports have been there in Asian patients.

DIFFERENTIAL DIAGNOSES

Pilonidal disease
Sacrococcygeal teratoma
Lipoma
Neurogenic tumor
Soft tissue sarcoma
Metastases

CONCLUSION

Subcutaneous myxopapillary ependymoma in sacrococcygeal region is extremely rare tumor with a tendency to affect children and adolescents. MRI is the imaging modality of choice for the detection, grading, and staging of this condition. Important treatment consideration is complete surgical excision at the initial operation. Long-term and close follow-up is required because this tumor may recur locally or become metastatic.

TEACHING POINTS

1. Although subcutaneous myxopapillary ependymoma is extremely rare entity but based on its sacrococcygeal location and characteristics seen on MRI, it should be considered as an important differential diagnosis and evaluated further.
2. Long-term post-operative, follow-up is suggested for evaluation of metastases.

MCQs

1. Which of the followings characterize subcutaneous myxopapillary ependymoma?
   a. World Health Organization grade II tumor
   b. Tends to affect children and adolescents
   c. Complete surgical excision is recommended treatment approach
   d. All the above

Answer Key: d

2. MRI findings in case of myxopapillary ependymoma include:
   a) Intermediate/high T2 signal intensity lesion
   b) Intense post-contrast enhancement
   c) Well-defined non-enhancing fibrous capsule with multiple internal non-enhancing fibrous septae
   d) All the above

Answer Key: d

3. Which of the following is true regarding myxopapillary ependymoma?
   a. Slow growing tumor
   b. No recurrence
   c. No distal spread
   d. Very high incidence

Answer Key: a

Ethical approval

Institutional Review Board approval is not required/waived-off.

Declaration of patient consent

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

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

Use of artificial intelligence (AI)-assisted technology for manuscript preparation

The authors confirm that there was no use of artificial intelligence (AI)-assisted technology for assisting in the writing or editing of the manuscript and no images were manipulated using AI.

REFERENCES

1. Helwig EB, Stern JB. Subcutaneous sacrococcygeal myxopapillary ependymoma. A clinicopathologic study of 32 cases. Am J Clin Pathol 1984;81:156-61.
2. Sonneland PR, Scheithauer BW, Onofrio BM. Myxopapillary ependymoma. A clinicopathologic and immunocytochemical study of 77 cases. Cancer 1985;56:883-93.
3. Louis DN, Perry A, Wesseling P, Brat DJ, Cree IA, Figarella-Branger D, *et al*. The 2021 WHO classification of tumours of the central nervous system: A summary. Neuro Oncol 2021;23:1231-51.
4. Ramkumar S, Wanniang CA, Wahlang AR, 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 2021;13:e14931.
5. Fujimori T, Iwasaki M, Nagamoto Y, Kashii M, Sakaura H, Yoshikawa H. Extraneural metastasis of ependymoma in the cauda equina. Global Spine J 2013;3:33-40.
6. Kelly A, Nally D, Crowther S, Kavanagh D. Subcutaneous sacrococcygeal myxopapillary ependymoma misdiagnosed as pilonidal disease. BMJ Case Rep 2020;13:e231639.
7. Johnson JM, Jessurun J, Leonard A. Sacrococcygeal ependymoma: Case report and review of the literature. J Pediatr Surg 1999;34:1405-7.
8. 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. Br J Plast Surg 2002;55:80-8.
9. Miralbell R, Louis DN, O'Keeffe D, Rosenberg AE, Suit HD. Metastatic ependymoma of the sacrum. Cancer 1990;65:2353-5.

How to cite this article: Dhandhalya CV, Chudasama SL, Pansuriya HK. Imaging in sacrococcygeal subcutaneous myxopapillary ependymoma: Sporadically reported from Asian population. Case Rep Clin Radiol. 2024;2:124-7. doi: 10.25259/CRCR_74_2023