jmr Junior Medical Research

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Case Report

Xanthogranuloma of the sellar region in a female patient presenting with pituitary dysfunction: A case report and review of literature.

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Publication Data:
Submitted: December 27,2024
Accepted: February12,2025
Online: April 30,2025

This article was subject to full peer-review.

Abstract

Xanthogranuloma of the sellar region is an extremely rare lesion. It represents approximately 1.9% of all sellar and parasellar region tumors with only eighty-four reported cases in the literature. The preoperative diagnosis is difficult due to the lack of clinical and radiological specificities. We herein report A new case of sellar Xanthogranuloma described in a young Tunisian patient.

Keywords
Craniopharyngioma; Magnetic Resonance Imaging; surgery; Xanthogranuloma.

Introduction

Xanthogranuloma of the sellar region is an extremely rare brain tumor. This lesion arises frequently in the middle ear, petrous apex, paranasal sinuses, choroid plexus and mostly located in the trigone of the lateral ventricle [1]. It represents about 1.9% of all tumors of the sellar region with only less than ninety previously described cases. These tumors were underdiagnosed and usually confused with xanthogranulomatous reaction of Craniopharyngioma or Rathke's cleft cyst [2].
Diagnosis is challenging because of the absence of typical clinical and radiological features for these tumors. Our aim was to report Xanthogranuloma management specificities according to the SCARE 2020 criteria.

Observation

We report the case of a 28-year-old female with a history of delayed puberty and pituitary insufficiency for seven years. the patient complained of gradual decrease in visual acuity of the right eye. Neurological examination showed no neurological deficits. Ophthalmological examination reveals visual acuity of 1/10 on the right side and 10/10 on the left side without any disturbance of the visual fields. Fundus examination showed optic atrophy on the right and papillary pallor on the left. Endocrinological tests were normal. Brain MRI showed well-defined 40X35X20 mm sellar and supra sellar tumoral lesion with mixed solid and cystic components. This lesion was of ovoid shape, presenting hyperintense signal on both T1- and T2-weighted sequences and displacing the optic chiasm (figure 1). Brain CT scan reveals the presence of calcifications located around the periphery of the lesion. Surgery was indicated. A complete resection was performed using a fronto-pterional approach. The histopathological examination, based on the presence of fibrous tissue with abundant cholesterol clefts, foamy macrophages, and hemosiderin deposits confirmed the diagnosis of Xanthogranuloma (figure2). Postoperative course was uneventful. The MRI follow up at 3 months did not show any tumor recurrence.

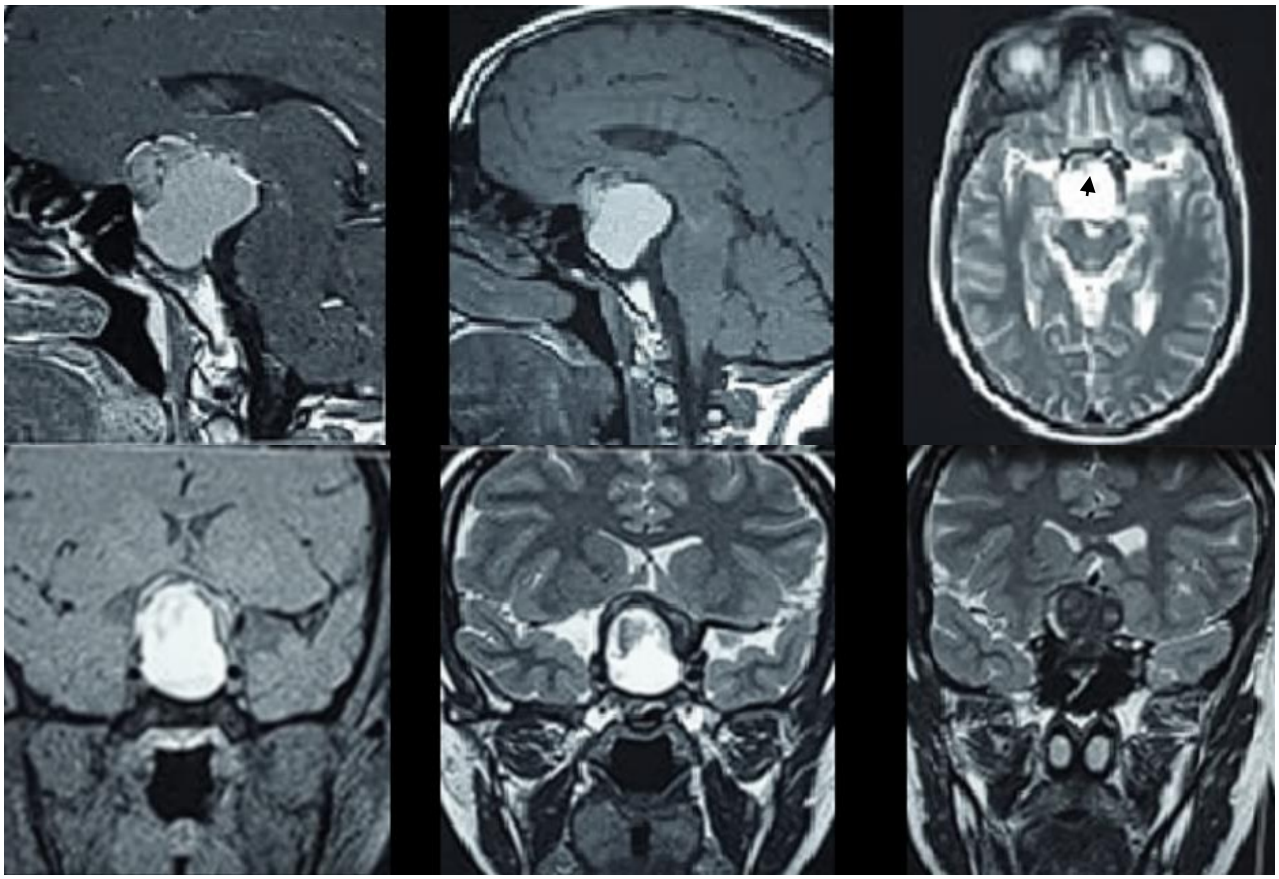

Figure 1: Preoperative sagittal T1-weighted magnetic resonance showing a an ovoid sellar and supra sellar tumor measuring 40*35*20 mm in diameter with solid and cystic components. The lesion presents a hyperintense signal on both T1- and T2-weighted sequences. The tumor compressed the pituitary gland and the optic chiasma upward.

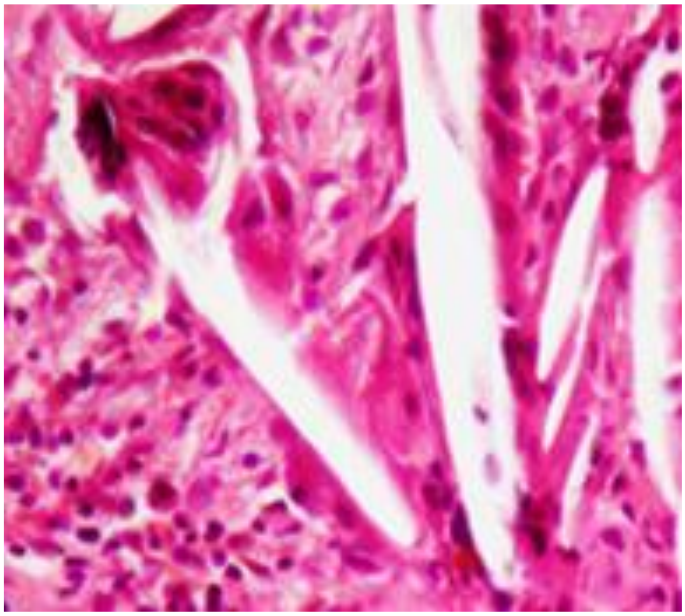

Figure 2: histopathological exam showed cholesterol cleft, chronic inflammatory cells, and collagenous fibrous tissue.

Citation: Mechergui H, Bouali S, Slimane A, Farhat S, Abderrahman Kh, Kallel J. Xanthogranuloma of the sellar region in a female patient presenting with pituitary dysfunction: A case report and review of literature. Jr. med. res. 2025; 7(1):9-11.
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Discussion

Xanthogranuloma of the sellar region is a rare entity that mostly presents in children, adolescents, and young adults [3]. Annual incidence approximates 0.05 per million inhabitants. Tumors occur in the choroid plexus of the lateral ventricles, and rarely in the sellar and parasellar region [4]. Xanthogranulomas were distinguished from craniopharyngiomas first in 1999 [5]. Sellar Xanthogranulomas are characterized by the presence of cholesterol crystals, hemosiderin deposits, lymphocytic infiltrates, macrophage accumulation, fibrous proliferation, and sparse clusters of epithelial cells [6]. However, they are still frequently misdiagnosed as craniopharyngiomas due to their macroscopic intraoperative features and the occasional presence of epithelial components. Clinical symptoms are of wide range and remain nonspecific. Xanthogranulomas may be responsible of several neurological and endocrine disorders such as visual impairment due to optic chiasm compression, panhypopituitarism, diabetes insipidus of central origin, and obstructive hydrocephalus [7]. Magnetic resonance imaging (MRI) is crucial step in the management of this entity. However, features are rarely specific. Cholesterol crystals are observed hyperintense on T1 weighted image and hypointense on T2. Hemosiderin cysts are iso or hyperintense on T1 sequence and hyperintense on T2. Fibrosis is hypointense on T1 and T2 images. After gadolinium administration, enhancement is usually heterogeneous [8].
Surgical resection is necessary and reduce mass effect neurological consequences. The evolution of the hormonal deficiency is variable. In our case, the patient's hormonal deficits did not improve, and she resumed her endocrine replacement therapy. The histopathological examination of the excised specimen confirms the Xanthogranuloma diagnosis in most of the cases.

Conflict of Interest: None

References

[1] Guerrero-Pérez F, Marengo AP, Vilarrasa N, Vidal N, Ruiz-Roig N, Sánchez-Fernández JJ, Torres-Díaz A, Sanmillán JL, Pérez-Maraver M, Villabona C. Xanthogranuloma of the sellar region : A systematic review. Hormones (Athens). 2023 Jun ;22(2) :199-210.
[2] Lozovanu V, Georgescu CE, Florescu LM, Georgiu C, Silaghi H, Fratea A, Silaghi CA. Xanthogranuloma of the Sellar Region: A Comprehensive Review of Neuroimaging in a Rare Inflammatory Entity. J Pers Med. 2022 Jun 8;12(6):943.
[3] Alharbi A, Alkhaibary A, Alaglan A, Khairy S, Alkhunaizi Z, AlSufiani F, Alassiri AH, Alkhani A. Sellar xanthogranuloma: A diagnostic challenge. Surg Neurol Int. 2022 Mar 4; 13:76.
[4] Cheng D, Yang F, Li Z, Qv F, Liu W. Juvenile Xanthogranuloma of the Sellar Region with a 5-Year Medical History: Case Report and Literature Review. Pediatr Neurosurg. 2021;56(5):440-447.
[5] La Rocca G, Rigante M, Gessi M, D'Alessandris QG, Auricchio AM, Chiloiro S, De Marinis L, Lauretti L. Xanthogranuloma of the sellar region: A rare tumor. Case illustration and literature review. J Clin Neurosci. 2019 Jan; 59:318-324.
[6] Rios IS, Silva DR, Goncalves PE, Souza Junior JF, Gurgel RQ, Oliveira AMP. Juvenile xanthogranuloma as a differential diagnosis for sellar and suprasellar lesions in child: Case report and review of literature. Neurochirurgie. 2023 Sep;69(5):101472.
[7] Kobayashi M, Yagasaki H, Kobayashi K, Ogiwara M, Kinouchi H, Sugita K. Xanthogranuloma of the sellar region accompanied by growth hormone deficiency: case report and literature review. J Pediatr Endocrinol Metab. 2018 Oct 25;31(10):1161-1164.
[8] Gurcay AG, Gurcan O, Kazanci A, Bozkurt I, Senturk S, Ferat M, Turkoglu OF, Beskonakli E, Orhun Yavuz HS. Xanthogranuloma of the sellar region. Neurol India. 2016 Sep-Oct;64(5):1075-9.
[9] Shao X, Wang C, Min J. Xanthogranuloma of the sellar region: A case report. Medicine (Baltimore). 2020 Oct 2;99(40): e22619.
[10] Dai CX, Guo XS, Liu XH, Bao XJ, Feng M, Zhong DR, Ma WB, Wang RZ, Yao Y. Xanthogranuloma of the Sellar Region. Chin Med J (Engl). 2017 Jan 20;130(2):249-250.

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