



Case Report

Sellar xanthogranuloma: A diagnostic challenge

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Received : 27 November 2021

Accepted : 12 January 2022

Published : 04 March 2022

DOI

10.25259/SNI_1180_2021

Quick Response Code:



ABSTRACT

Background: Sellar xanthogranulomas are rare intracranial lesions comprising <1% of all sellar lesions. They were described as a separate entity by the World Health Organization in 2000. Because of the paucity of sellar xanthogranuloma cases reported in the literature, they remain a diagnostic challenge with indefinite origin, clinical course, and outcome. The present study reports a case of sellar xanthogranuloma describing the clinical presentation, radiological/pathological characteristics, and outcome.

Case Description: A 43-year-old female, known to have diabetes, hypothyroidism, and polycystic ovarian syndrome, presented with a 2-week history of sudden right-sided facial deviation, periorbital pain, and moderate-intensity headache. The patient also reported amenorrhea not improving with polycystic ovarian syndrome treatment. Neurologic examination showed bilateral visual field defects and impaired visual acuity. Computed tomography scan, without contrast, revealed a hypodense sellar lesion with areas of hyperdensity. Magnetic resonance imaging showed a well-defined sellar lesion, exhibiting high signals on T1-weighted and T2-weighted images. The patient underwent microscopic trans-nasal trans-sphenoidal excision of the lesion. Histological sections of the sellar lesion revealed fibrous connective tissue with chronic inflammatory cells and cholesterol clefts, suggestive of xanthogranuloma. The patient is currently followed up at neurosurgery, endocrinology, and ophthalmology clinics with periodic laboratory/radiological investigations.

Conclusion: Sellar xanthogranulomas remain rare intracranial lesions with few cases reported in the literature. Patients mostly present with severe hypopituitarism and visual dysfunction. They show no characteristic radiological features. The diagnosis is confirmed histopathologically, and the prognosis is generally favorable.

Keywords: Adenohypophysis, Cholesterol, Fibrous, Foamy, Granuloma, Pituitary

INTRODUCTION

Sellar xanthogranulomas, also known as cholesterol granulomas, are rare brain lesions comprising 0.6% of all sellar tumors.^[9] In 2000, sellar xanthogranulomas were considered an independent clinical entity by the World Health Organization.^[10] Patients classically present with hypopituitarism and/or visual dysfunction.^[8] There are no pathognomonic radiological features of xanthogranulomas.^[1] Histologically, they consist of cholesterol clefts, multinucleated giant cells, lymphocytic infiltrate, foamy macrophages, fibrous proliferation, hemosiderin deposits, and necrotic debris.^[8] Differential diagnoses of xanthogranulomas include

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craniopharyngioma, Rathke's cleft cyst, and hemorrhagic pituitary adenoma.^[1] The more common differential diagnoses are initially considered due to the rarity of sellar xanthogranuloma, rendering the diagnosis delayed.

Because of the paucity of sellar xanthogranuloma cases reported in the literature, they remain a diagnostic challenge with indefinite origin, clinical course, and outcome. The present study reports a case of sellar xanthogranuloma by describing the clinical presentation, radiological/pathological characteristics, and outcome.

CASE DESCRIPTION

Clinical presentation

A 43-year-old female was known to have diabetes, hypothyroidism, and polycystic ovarian syndrome. The patient was in her usual state of health until 2 weeks before her presentation when she developed a sudden right-sided facial deviation, periorbital pain, and moderate-intensity headache. The patient acknowledged chronic history of mild bifrontal headache for the past 3 years, for which she did not seek medical attention. Furthermore, the patient reported amenorrhea for the same duration, not improving with polycystic ovarian syndrome treatment. On further inquiry, the patient reported difficulties in visualizing objects located in her peripheral fields. There was no history of galactorrhea, cold/heat intolerance, weight gain/loss, or hirsutism.

Physical examination

The patient was alert and oriented with a Glasgow Coma Scale of 15/15. The pupils were 3 mm reactive to light and accommodation bilaterally. The extraocular muscles movement was intact with no ptosis. The color vision test was normal. The corneas and anterior chambers were clear. Fundoscopy revealed healthy discs and maculae bilaterally. The 24-2 Humphrey test revealed bilateral visual field defects. The visual acuity was impaired bilaterally (right eye: 20/30 and left eye: 20/50). Otherwise, the patient was neurologically intact.

Neuroradiological imaging

Computed tomography (CT) scan without contrast revealed a hypodense sellar lesion with areas of hyperdensity [Figures 1a and b]. Pituitary magnetic resonance image (MRI), with gadolinium administration, showed a well-defined sellar lesion, exhibiting high signals on T1-weighted images (T1WI) and T2-weighted images (T2WI) [Figures 1c-e]. Given the clinical data and radiological features, the presumptive differential diagnoses included pituitary apoplexy, cystic hemorrhagic macroadenoma, and complex hemorrhagic Rathke's cleft cyst.

Surgical intervention

Considering the brain CT and MRI findings, surgical resection of the lesion was performed to establish the diagnosis. The patient underwent microscopic trans-nasal trans-sphenoidal excision of the pituitary gland. Intraoperatively, the tumor was readily visualized. Multiple samples were obtained and sent for permanent histopathology. Most of the visualized parts of the tumor were removed. Grossly, the tumor consisted of multiple pieces of tan, soft-tissue measuring, in aggregate, 1 × 1 × 0.5 cm.

Histopathological findings

Histological sections of the sellar lesion revealed fibrous connective tissue with chronic inflammatory cells and cholesterol clefts. Scattered foreign body giant cells were noticed with focal hemosiderin deposits and foamy histiocytes [Figures 2a-c]. A fragment of normal pituitary adenohypophysis was also appreciated [Figure 2d]. Henceforth, a diagnosis of sellar xanthogranuloma was made.

Outcome and follow-up

The patient tolerated the surgery well without complications. She was kept in the intensive care unit for 2 days for close monitoring. Serum sodium, serum osmolality, and urine osmolality levels were all within normal limits. There was no clinical or laboratory evidence suggestive of diabetes insipidus. Apart from her preexisting hypothyroid state, the pituitary hormonal profile was within normal during her hospital stay. Brain CT scan, 1 day following surgery, confirmed resection of the lesion [Figure 3]. During hospitalization, no cerebrospinal fluid leakage was noted. The patient was discharged in a stable condition. One month following the discharge, the patient reported subjective improvement of her preexisting visual impairment. The patient has regular clinical follow-up at neurosurgery, endocrinology, and ophthalmology clinics with laboratory/radiological investigations.

DISCUSSION

Sellar xanthogranulomas are rare intracranial lesions with few cases reported on review of the literature. We hereby report an additional case of such uncommon sellar lesion encountered in our tertiary care center.

The origin of sellar xanthogranulomas is yet undetermined. Sellar xanthogranulomas were first considered as a variant of adamantinomatous craniopharyngioma.^[2] Shirataki *et al.*^[11] reported the first histological description of "pure cholesterol xanthomatous reaction" in the sellar region in 1988. Paulus *et al.*^[8] first described sellar xanthogranulomas as a separate

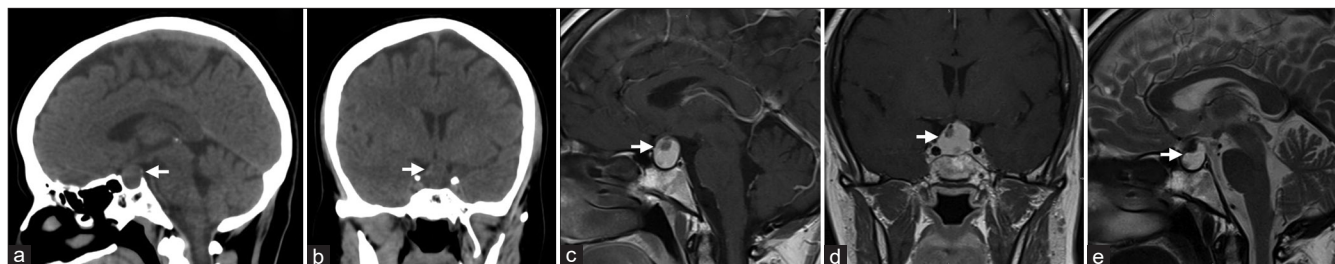


Figure 1: (a and b) Sagittal and coronal brain CT without contrast showing a hypodense lesion with areas of hyperdensities (Arrow). (c and d) Sagittal and coronal T1-weighted images with contrast of the pituitary gland. (e) Sagittal T2-weighted image. (c-e) The images demonstrate a well-defined, rounded sellar mass which exhibit a high signal on T1- and T2-weighted images (Arrow). The lesion causes deviation of the pituitary stalk to the left side and abuts the optic chiasm. It measures $19 \times 20 \times 12$ mm in anteroposterior, transverse, and craniocaudal diameters.

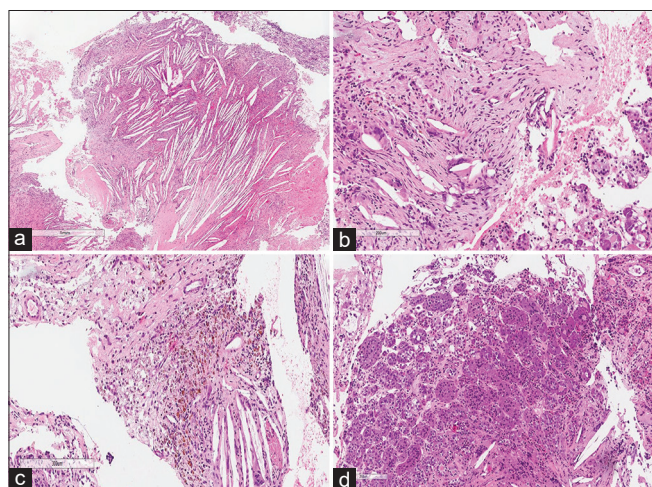


Figure 2: (a) Cholesterol clefts (H&E, original magnification $\times 100$). (b) Foreign body giant cells between the cholesterol clefts (H&E, original magnification $\times 200$). (c) Hemosiderin deposits (H&E, original magnification $\times 200$). (d) Pituitary adenohypophysis (H&E, original magnification $\times 200$).

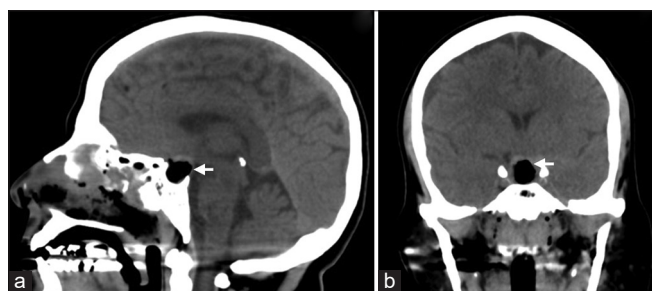


Figure 3: (a and b) Postoperative sagittal and coronal brain CT without contrast demonstrating expected postoperative changes in the form of opacification of the sinonasal cavity as well as fragmented pockets of air foci, along the hemorrhagic densities, within the surgical cavity (Arrow).

entity different from craniopharyngioma.^[8] They found 37 tumors with a predominant xanthogranulomatous component among 110 cases of craniopharyngioma.^[8] Of which, three cases showed features of adamantinomatous

craniopharyngioma, and 13 cases showed nonadamantinomatous epithelium.^[8]

Xanthogranulomas with squamous epithelium and calcifications have been reported to arise from adamantinomatous craniopharyngioma.^[9,11] Le *et al.*^[6] reported that xanthomatous change was observed in 46% of Rathke's cleft cysts. Amano *et al.*^[2] reported that xanthogranulomatous inflammation was observed in seven cases of 123 cases of Rathke's cleft cyst and 51 cases of craniopharyngioma.

Pituitary xanthogranulomas are radiologically indistinct and show mixed intensities due to the complex histologic components of the lesions. It has been speculated that hemosiderin deposits show hypointense signals on T2WI.^[1] Cholesterol clefts show high and low T1WI and T2WI signals, respectively.^[10] Fluid components within the cystic lesions appear as high T2WI signal intensities.^[10] The presence of extensive fibrosis and hemorrhage can show low T1WI and T2WI signal intensities.^[10] Ved *et al.*^[12] reported that more than 80% of xanthogranuloma cases showed high T1WI signal intensities. Although calcifications are rare in xanthogranuloma, they have been reported in the literature.^[1,3] In the present case, the sellar lesion showed high T1WI and T2WI signals with no areas of calcification.

Surgical resection remains the mainstay of treatment of sellar xanthogranulomas with gross total resection as the gold standard.^[12] In general, endoscopic endonasal approach is superior to transcranial approaches.^[7] Visual dysfunction was reported to successfully subside after effective surgical decompression.^[13] In our case, the patient reported improvement in her visual symptoms on follow-up in the clinic. Preoperative diabetes insipidus, a common presentation of pituitary xanthogranulomas, takes a relatively long time to recover postoperatively.^[12] In our case, the patient did not present with diabetes insipidus. Recurrence is rare after total or subtotal resection.^[4,5]

Considering the rarity of sellar xanthogranuloma, the clinical presentation, and radiological features of the present case, other differential diagnoses of sellar lesions were initially considered. The patient underwent near total resection of

the lesion due to its adherence to the diaphragma sellae. The diagnosis of sellar xanthogranuloma was confirmed histopathologically, and the patient had an excellent outcome.

CONCLUSION

Sellar xanthogranulomas remain rare intracranial lesions with few cases reported in the literature. Patients mostly present with severe hypopituitarism and visual dysfunction. They show no characteristic radiological features. The diagnosis is confirmed histopathologically, and the prognosis is generally favorable.

Acknowledgment

The authors would like to express their gratitude to King Abdullah International Medical Research Center, Ministry of National Guard - Health Affairs, Riyadh, Saudi Arabia for approving the study. The assigned protocol number for approval is NR21R/358/09.

Authors' contributions

Ahoud Alharbi: Writing – Original Draft, Writing – Review and Editing. Ali Alkhaibary: Conceptualization, Investigations – Radiological and Pathological images, Writing – Original Draft, Writing – Review and Editing. Abeer Alaglan: Conceptualization, Investigations – Radiological and Pathological images, Writing – Original Draft, Writing – Review and Editing. Sami Khairy: Conceptualization, Supervision, Writing – Review and Editing. Zahra Alkhunaizi: Investigations – Pathological images, Microscopic Description, Writing – Review and Editing. Fahd AlSufiani: Conceptualization, Investigation – Microscopic Description, Supervision, Writing – Review and Editing. Ali H. Alassiri: Conceptualization, Investigation – Microscopic Description, Supervision, Writing – Review and Editing. Ahmed Alkhani: Surgical Management, Supervision, Writing – Review and Editing. All authors have critically reviewed and approved the final version of the manuscript.

Declaration of patient consent

Institutional Review Board (IRB) permission obtained for the study.

Financial support and sponsorship

Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Agarwal A, Agarwal K, Lee HK. Xanthogranuloma of the sellar region: A case report. *Neuroradiol J* 2012;25:181-4.
2. Amano K, Kubo O, Komori T, Tanaka M, Kawamata T, Hori T, et al. Clinicopathological features of sellar region xanthogranuloma: Correlation with Rathke's cleft cyst. *Brain Tumor Pathol* 2013;30:233-41.
3. Fujio S, Takajo T, Kinoshita Y, Hanaya R, Arimura H, Sugata J, et al. Sellar xanthogranuloma: A quest based on nine cases assessed with an anterior pituitary provocation test. *World Neurosurg* 2019;130:e150-9.
4. Hernández-Estrada RA, Kshetry VR, Vogel AN, Curtis MT, Evans JJ. Cholesterol granulomas presenting as sellar masses: A similar, but clinically distinct entity from craniopharyngioma and Rathke's cleft cyst. *Pituitary* 2017;20:325-32.
5. Kleinschmidt-DeMasters BK, Lillehei KO, Hankinson TC. Review of xanthomatous lesions of the sella. *Brain Pathol* 2017;27:377-95.
6. Le BH, Towfighi J, Kapadia SB, Lopes MB. Comparative immunohistochemical assessment of craniopharyngioma and related lesions. *Endocr Pathol* 2007;18:23.
7. Nishiuchi T, Murao K, Imachi H, Kushida Y, Haba R, Kawai N, et al. Xanthogranuloma of the intrasellar region presenting in pituitary dysfunction: A case report. *J Med Case Rep* 2012;6:1-7.
8. Paulus W, Honegger J, Keyvani K, Fahlbusch R. Xanthogranuloma of the sellar region: A clinicopathological entity different from adamantinomatous craniopharyngioma. *Acta Neuropathol* 1999;97:377-82.
9. Rahmani R, Sukumaran M, Donaldson AM, Akselrod O, Lavi E, Schwartz TH. Parasellar xanthogranulomas. *J Neurosurg* 2015;122:812-7.
10. Shao X, Wang C, Min J. Xanthogranuloma of the sellar region: A case report. *Medicine (Baltimore)* 2020;99:e22619.
11. Shirataki K, Okada S, Matsumoto S. Histopathological study of the "cholesterol granuloma reaction" in the sellar and juxtaseellar tumors. *No To Shinkei* 1988;40:133-9.
12. Ved R, Logier N, Leach P, Davies JS, Hayhurst C. Pituitary xanthogranulomas: Clinical features, radiological appearances and post-operative outcomes. *Pituitary* 2018;21:256-65.
13. Yang B, Yang C, Fang J, Li G, Wang J, Jia G, et al. Clinicoradiologic features and surgical outcomes of sellar xanthogranulomas: A single-center 10-year experience. *World Neurosurg* 2017;99:439-7.

How to cite this article: Alharbi A, Alkhaibary A, Alaglan A, Khairy S, Alkhunaizi Z, AlSufiani F, et al. Sellar xanthogranuloma: A diagnostic challenge. *Surg Neurol Int* 2022;13:76.