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XANTHOGRANULOMA OF THE SELLAR REGION IN A PATIENT WITH SARCOIDOSIS

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SUMMARY – Xanthogranuloma of the sellar region is a very rare brain tumor with favorable prognosis and without reported relapses of purely xanthogranulomatous lesion after complete resection. A case is presented of a 40-year-old male diagnosed with and treated for sarcoidosis, complaining of headache, photophobia and loss of libido. Physical examination revealed generally scarce hairiness, while laboratory investigations showed panhypopituitarism. Expansive sellar and suprasellar mass compressing the floor of the third ventricle and optical chiasm was confirmed by cranial multi-slice computerized tomography (MSCT). Complete resection of the tumor mass using trans-sphenoidal approach was performed. Histopathologic analysis revealed cholesterol clefts, sparse lymphoplasmacellular infiltrates, macrophages, siderophages and foreign body giant cells around cholesterol clefts confirming the diagnosis of xanthogranuloma of the sellar region. Since preoperative diagnosis of xanthogranuloma is very difficult, therapeutic algorithm does not differ from other expansive lesions of the sellar region, but pituitary involvement should always be considered in patient with sarcoidosis since therapeutic management is non-surgical. Follow up MSCT imaging after 6 months revealed a solid, contrast-enhanced mass at the posterior base of the sella.

Key words: *Pituithry neoplasms – diagnosis, surgery; Sella turcica – pathology; Xantogranuloma – diagnosis, surgery; Sarcoidosis – complications; Case report*

Introduction

Xanthogranuloma of the sellar region is a very rare brain tumor[1-3]. The first described series of 37 cases were reported by Paulus *et al.* in 1999, when xanthogranuloma of the sellar region was proposed as a distinct tumor, separate from craniopharyngeoma[1]. The updated World Health Organization classification of brain tumors from 2000 has accepted xanthogranuloma as a separate entity[4]. Distinct characteristics of xanthogranuloma of the sella have been described previously[1] and are summarized in Table 1. Generally, xanthogranuloma of the sella has a favorable prognosis with no reported relapses of purely xanthogranulomatous lesion after complete resection[1,3]. Herein, we report on a patient with a typical presentation of xanthogranuloma of the sellar region, in whom follow up revealed the first possible case of relapse after complete resection.

Case Report

A 40-year-old male patient presented complaining of a 5-year history of headache, photophobia, and loss of libido. Two years before, the patient had been diagnosed with sarcoidosis and received steroid therapy at the time of admission. Clinical examination revealed no signs of cranial nerve palsy or other neurologic deficits. Other physical findings were unremarkable except for generally scarce hairiness. Further laboratory

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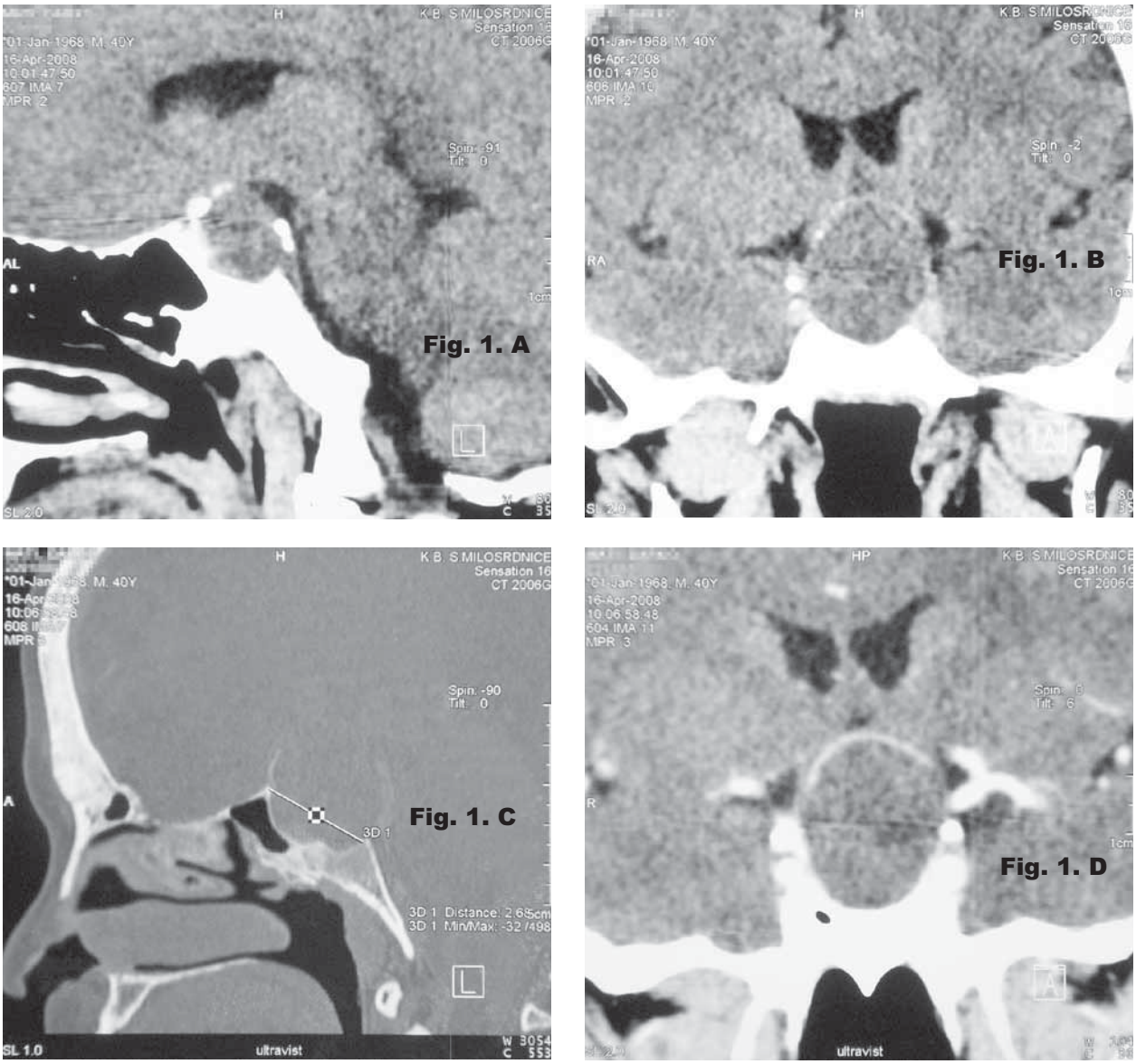
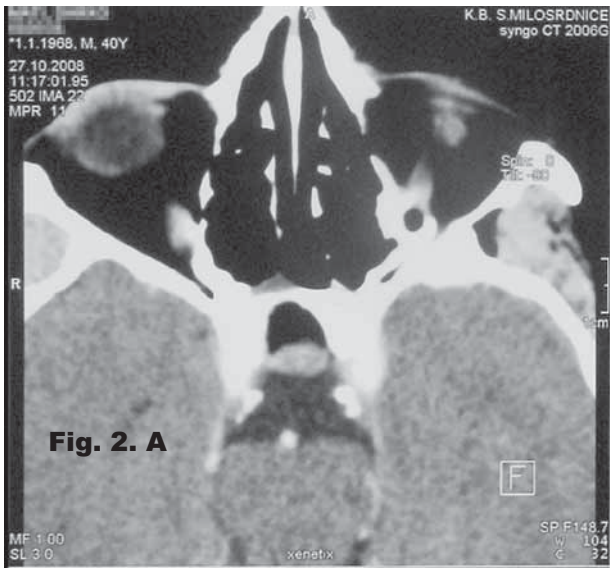
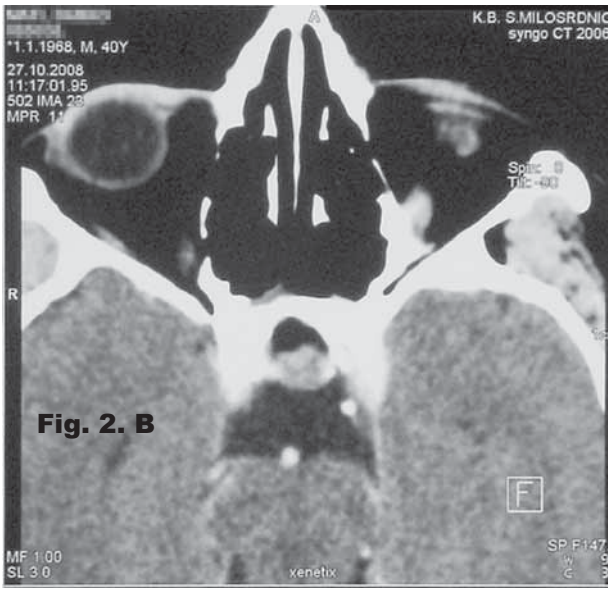
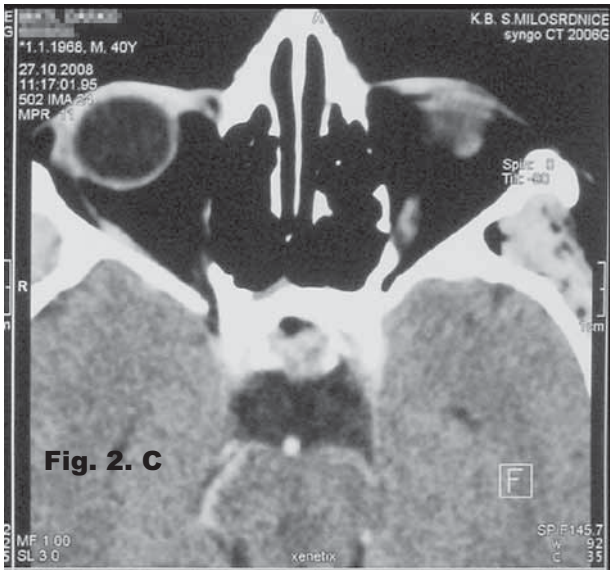


Fig. 1. Preoperative MSCT of the sellar region revealing expansive intrasellar and suprasellar mass in sagittal and frontal views (A and B); sella turcica is enlarged (C); margins of the expansive mass are contrast enhanced (D).

examinations were within the normal limits except for endocrinologic results that showed panhypopituitarism. Cranial MSCT scan revealed enlarged sella turcica and a 2.5x2 cm expansive sellar and suprasellar mass compressing the floor of the third ventricle and optic chiasm. The lesion showed a hypodense cystic center and hyperdense, contrast-enhanced anterior segment (Fig. 1). The tumor was completely removed using a transsphenoidal approach under a presumptive diagnosis of pituitary macroadenoma. The cystic tumor contained yellowish fluid which was drained, and a small, solid sample of tumor wall tissue was obtained for histologic analysis. Histopathologic analysis of frozen and permanent sections revealed several tissue specimens measuring up to 0.7 cm in greatest dimension, composed of cholesterol clefts, sparse lymphoplasmacellular infiltrates, macrophages, siderophages and foreign body giant cells around cholesterol clefts. Epithelial cells with peripheral palisading, keratohyaline granules and wet keratin were







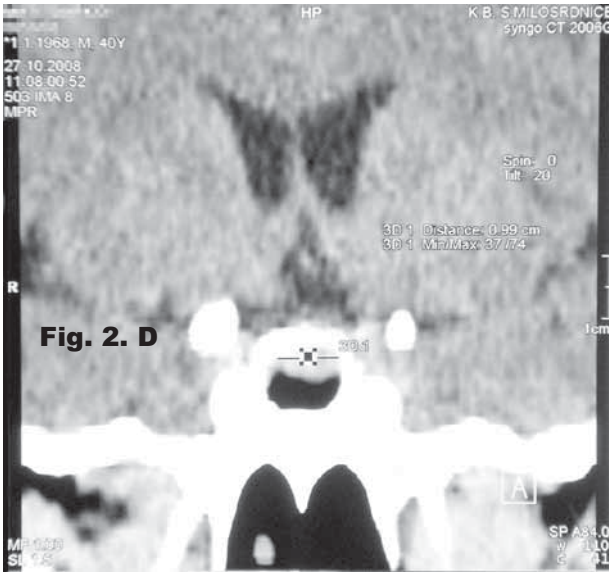


Fig. 2. Postoperative MSCT of the sellar region revealing residual tumor mass in transverse (A and B) and frontal view (C).

absent, favoring the histopathologic diagnosis of sellar region xanthogranuloma.

Six-month postoperative follow up showed improved patient's condition and absence of previous symptoms but requiring continuous medical supplementation for pituitary insufficiency. Follow up MSCT imaging at 6 months revealed a solid, contrast-enhanced mass at the posterior base of the sella (Fig. 2).

Discussion

Differential diagnosis of xanthogranuloma of the sellar region has been described elsewhere[2,3]. Several demographic and clinical characteristics could facilitate suspicion of xanthogranuloma in patients presenting with expansive sellar mass[1]. Adolescent or young adult, marked endocrine deficits and intrasellar involvement are common characteristics of a patient with sellar xanthogranuloma despite several reported

Table 1. Characteristics of xanthogranuloma of the sellar region

Age at onset	20-40 years (adolescents and young adults)[1] Cases in childhood and fifth or sixth decade have also been reported[1,2,5,6]
Anatomic site	Intrasellar region
Symptoms	Endocrinologic symptoms (common) Visual changes due to compression of optic pathways Obstructive hydrocephalus (rare)[3]
Prognosis	Favorable outcome
Histologic signs	Cholesterol-cleft granulomas Hemosiderin deposits Always absent palisading of peripheral epithelial cells, keratohyaline granules and wet keratin
Radiological signs	Nonspecific, differentiation from other expansive processes of the sellar region is not possible

exceptions, especially considering patient age[3]. Since there are no typical radiological signs pathognomonic of xanthogranuloma, radiological differentiation from other diagnoses is not possible[2]. Furthermore, recent reports suggest that an accurate preoperative diagnosis of xanthogranuloma of the sella, either clinical or radiological, is very difficult[3]. Since surgical intervention is necessary to correct the underlying endocrine problem and/or impairment of the visual pathway, therapeutic algorithm does not differ from other expansive lesions[2]. Nevertheless, differential diagnosis, apart from the conditions proposed, should be broadened in patients presenting with expansive sellar mass and sarcoidosis. In spite of rare pituitary presentation of sarcoidosis, this possibility should not be neglected since therapeutic management differs dramatically[8].

All reported cases of xanthogranuloma of the sella had favorable overall outcome with no reported relapses[2,3,5-7], including the first large series described by Paulus *et al.*, when no pure xanthogranulomatous lesion recurred after complete resection. In the study conducted by Paulus *et al.*, both of the two recurrences reported (frequency of 7.1%) had additional small portions corresponding to calcifying odontogenic cyst on histologic examination[1]. Since reports on xanthogranuloma are rare, it is important to report all cases to help reveal the true nature and clinical course of the disease. In our patient, follow up imaging showed the possible residue or relapse and further follow up is obligatory.

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Sažetak

KSANTOGRANULOM SELARNOG PODRUČJA U BOLESNIKA SA SARKOIDOZOM

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Ksantogranulom selarnog područja je vrlo rijedak tumor mozga s povoljnom prognozom i bez opisanih recidiva čisto ksantogranulomatozne lezije nakon potpunog odstranjenja. Prikazuje se slučaj 40-godišnjeg bolesnika s dijagnosticiranom i liječenom sarkoidozom koji se tužio na glavobolju, fotofobiju i gubitak libida. Fizikalni pregled je pokazao oskudnu dlakavost, dok su laboratorijske pretrage otkrile panhipopituitarizam. Ekspanzivnu selarnu i supraselarnu tvorbu koja je pritiskala dno trećeg ventrikla i optičkog hijazma potvrdila je višeslojna kompjutorizirana tomografija (MSCT). Tumorska tvorba je odstranjena u potpunosti transsfenoidnim pristupom. Histopatološka analiza je pokazala nakupine kolesterola, rijetke infiltrate limfoplazmatskih stanica, makrofage, siderofage i divovske stanice stranog tijela oko nakupina kolesterola, potvrđujući dijagnozu ksantogranuloma selarnog područja. Kako je prijeoperacijsku dijagnozu ksantogranuloma vrlo teško postaviti, terapijski algoritam se ne razlikuje od onoga za druge ekspanzivne lezije selarnog područja, no zahvaćenost hipofize treba uvijek imati na umu u bolesnika sa sarkoidozom, jer je liječenje neoperacijsko. Kontrolni MSCT nakon 6 mjeseci pokazao je čvrstu, kontrastom pojačanu tvorbu na stražnjoj bazi sele.

Ključne riječi: *Novotvorine hipofize – dijagnostika; Novotvorine hipofize – kirurgija; Sedlasta jama – patologija; Ksantogranulom – dijagnostika; Ksantogranulom – kirurgija; Sarkoidoza – komplikacije; Prikaz slučaja*