

Intra et parasellar chordoma – case report

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Abstract

A rare case of giant intra and parasellar chordoma, mimicking a pituitary prolactinoma is presented. The tumor involving pituitary fossa and surrounding structures was removed via a bifrontal approach. The pathology exhibited the typical histological patterns of a chordoma. The authors present a rare case of a giant sellar and parasellar chordoma and review the literature on that.

Keywords: chordoma, sella turcica, pituitary prolactinoma

Introduction

Chordomas represent 1% of all malignant bone tumors and 0.1 to 0.2% of intracranial neoplasms (6, 7). In Romania, the Pathology Department of the Bucharest Oncologic Institute has reported a low frequency for chordoma 0.12%. These embryonic tumors, first described by Luschka in 1856 are rare, slow-growing neoplasms with malignant potential, that arise from cell rests, particularly of the proximal and distal extremes of the notochord, characterized by osseous permeation, high local recurrence, local invasiveness (4-14). Their metastatic potential cannot be estimated by histological examination, except in cases of sarcomatous transformation (10). The most common

site are: 50% in the sacrococcygeal region – mean age 50 years, especially to men, 35% in the base of skull - in the midline, especially to children or young adults; 15% in vertebrae (7). Large, intra et parasellar chordomas resembling a pituitary prolactinoma are rare (12).

Case presentation

A 64 year old woman, without pathological antecedents, was admitted in our department for headache, progressive decrease of visual acuity of right eye, double vision installed over the past 2 months. On general and neurological examination we found decrease of visual acuity of right eye: VOD = 2/3 without correction, VOS = 1, the external rotation of the right eyeball was not possible, visual field examination disclosed bitemporal hemianopsia; fundus oculi: plane, well contoured papilla, with tortuous and dilated veins, thin arteries, macula without reflex on right eye; the light reflex of both pupils in direct & indirect responses was normal. No endocrinopathy was noted but serum pituitary hormone levels revealed high prolactin levels: 84 ng/ml, reference value 2.5–17, less basal cortisolemia 1,24 µg/dl, reference value 5–25, beta chorionic gonadotropin (hCG), alpha-fetoprotein (FP) and carcinoembryonic antigen (CEA) were

normal; diabetes insipidus investigation also was normal.

Radiography of the skull revealed enlargement and distortion of the sella, with osteolysis of its floor and invasion of the sphenoid bone. A high-resolution CT scan revealed a large, partly calcified intrasellar tumor, showing suprasellar extension and erosion of the sphenoid bone.

Cerebral MRI (Figure 1) demonstrated a sellar area tumor measured 28/32/34 mm, in its greatest diameter, with hypersignal in T1 and T2-weighted images, omogenous, with slight inside calcifications, polycyclic outlined. This tumor is elevating the pituitary gland and optic chiasm, posterior overpass clivus, is in contact with the basillary artery and on the right side invaded the latero-sellar compartment, without affecting the carotid artery. Therefore the carotid arteriography was normal.

A bifrontal microscopic approach was performed; a yellow encapsulated, soft with necrotic center tumor was discovered, extended intra, supra and right parasellar. The tumor was removed, the latero-sellar compartment too, the optic chiasm was decompressed as like as the pituitary stalk, less a small part of the tumor with tight adhesion to the pituitary stalk. Microcalcifications in the suprasellar and sellar portions were detected. Postoperative evolution was uneventfull confirmed by cerebral CT (Figure 2) and at 6-months follow-up examination after radiotherapy, the patient was asymptomatic, serum pituitary hormone levels revealed normal levels.

Histologic and immunohistochemistry tests are indicative for a tumoral chordoma. Pathology diagnosis was made on macroscopic criteria: lobulated fragments, tan-to-red, of various dimensions, soft consistency, sometimes kistic, diffuse borders, invading adjacent structures.

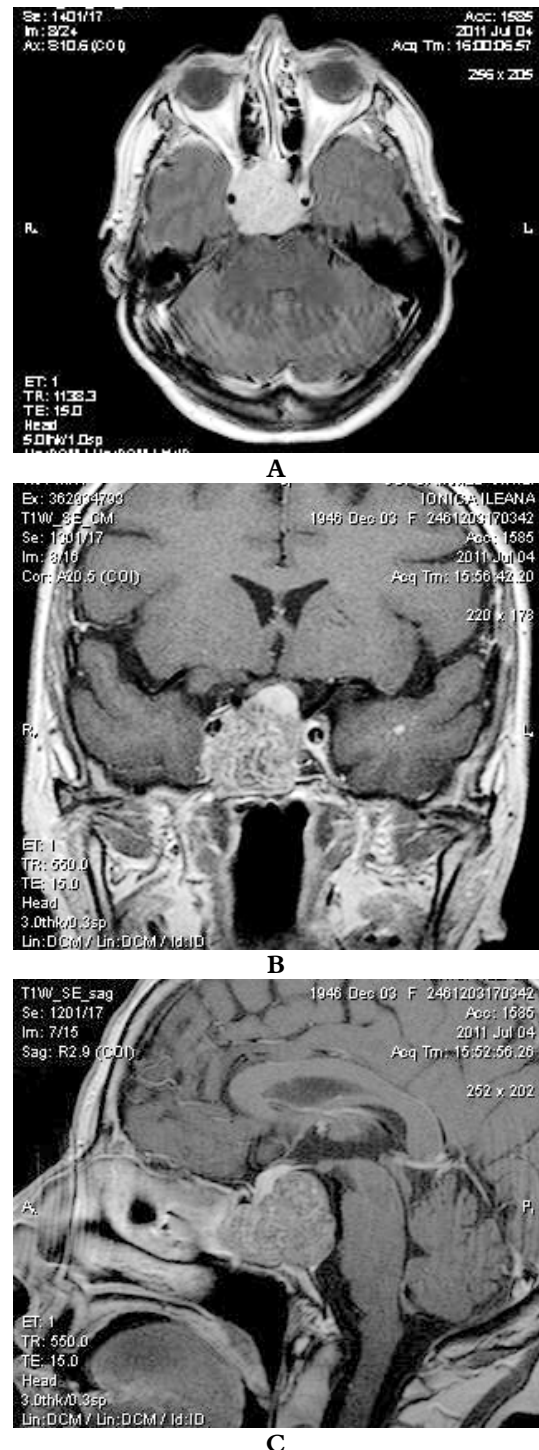


Figure 1 Cerebral MRI studies: axial (A), coronal (B) and sagittal (C) revealing a large mass occupying the pituitary fossa, elevating the pituitary gland and optic chiasm, and invading the right cavernous sinus

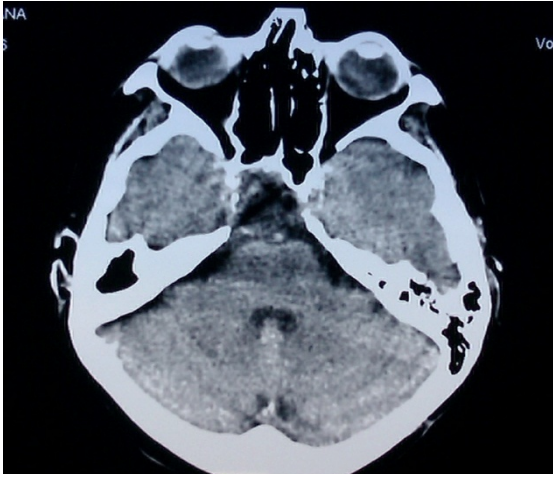


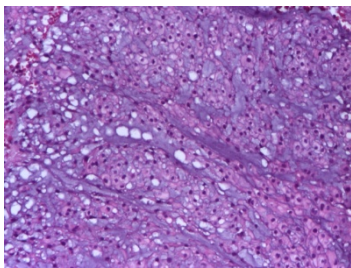
Figure 2 Postoperative CT-scan 1 month after operation

On section tumor is gelatinous, translucent with green with red-brune areas. Microscopic criteria (Figure 3): lobular pattern, more important in central part of the tumor, with different shapes, fibrous septa of variable thickness, more important at the tumor periphery, containing vessels. The arterial supply in the central area were very small, but necrosis was very often encountered. Smaller lobuli, perhaps younger, disposed at the periphery, contain oval, polygonal or prismatic cells, mimicking a carcinomatous aspect. This cells has important cytoplasm, homogenous, with small vacuolar areas containing glycogen and progressive tendency to coalescence. The nuclei are small, spheric – rarely with reniform shape, disposed central with prominent nucleoli. In the cells with

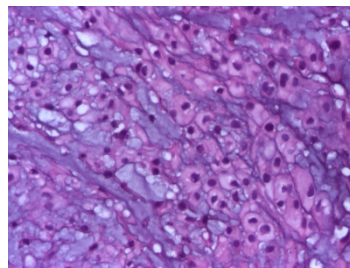
multiple vacuolar areas, nuclei has the tendency to picnosis.

In medium or big lobuli the proliferative cells were disposed circumferential and extended to the centrum part of the lobuli with a radial structure. In the central area, intracytoplasmatic vacuoles develop in volume and number; by vacuoles coalescence cells has the tendency to become “physaliphorous cells” with small nuclei disposed central or in the periphery (physaliphorous comes from the greek word physalis = bubble). Gigant, multinucleated cells are rare. Cytoplasmic vacuoles were smaller than those in the intercellular matrix. Another microscopic criteria was represented by an important mucoid stroma with glycogenic component (see the period-acid-Schiff method), where cells arranged in lobules are embedded. Very rare cells has pseudo-glandular aspects with lymphocytes at the periphery, partial limited by a capsula made by conjunctive tissue.

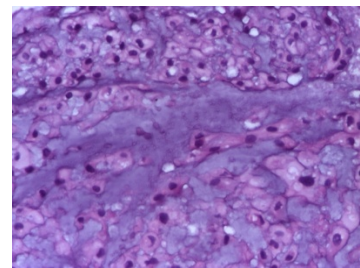
Immunocytochemical analysis of the chordoma cells showed focal positivity for vimentin, EMA. Negative immunoreactivity for CEA was noted. Ultrastructurally, the tumor was composed of large cells containing irregular nuclei with multiple indentations and prominent nucleoli. Analysis of DNA revealed a diploid histogram.



A



B



C

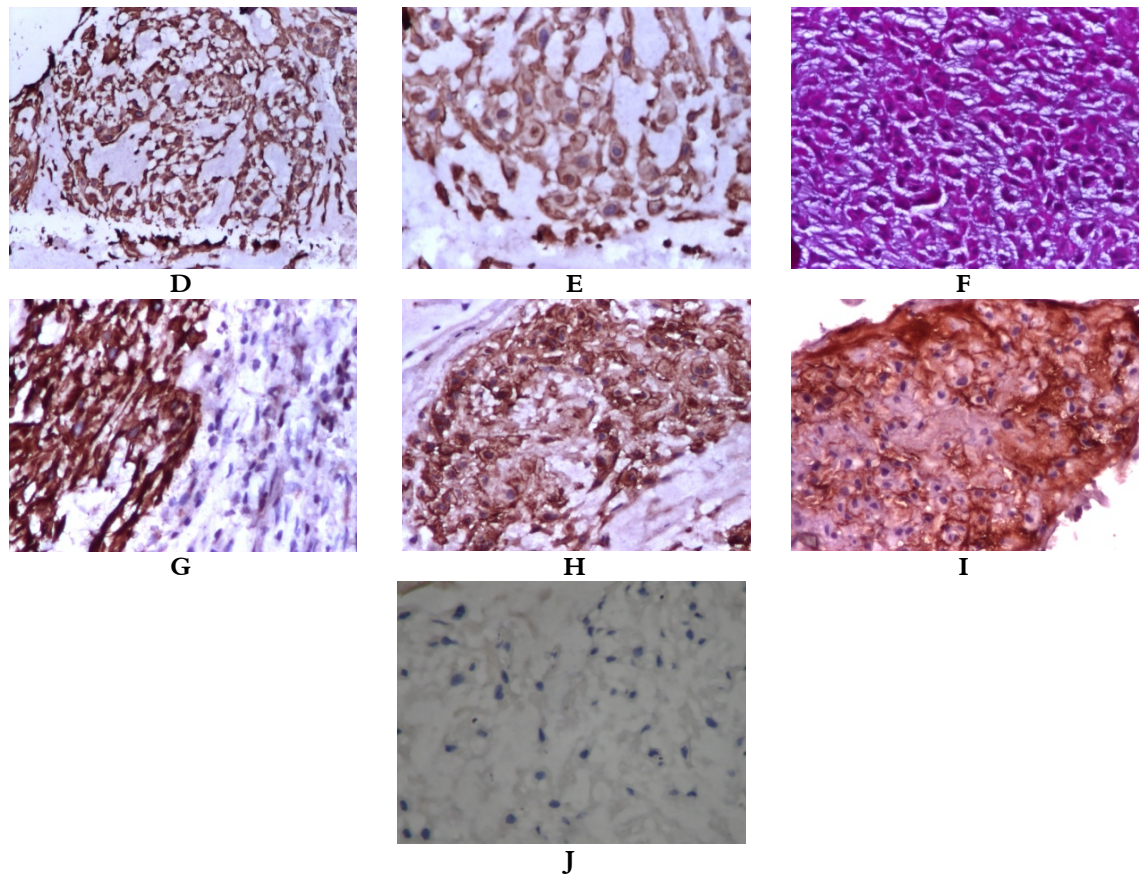


Figure 3 Chordoma: microscopic criteria and details for immunohistochemistry

- (A) chordoma HE - Ob.10 X: tumoral lobes, different shapes, separated by fibrotic septa, (B) detail: chordoma HE - Ob. 20 X: as vacuoles are bigger, the cells limit are estomped, (C) another detail of chordoma HE - Ob. 20 X, (D) VIM positive IHC coloration - Ob.10 X, (E) detail of VIM positive IHC coloration - Ob. 20 X, (F) PAS present as a marker of glycogen in mucilaginous substance expelled in extracellular space - Ob. 20 X, (G) detail of MNF (myelinated nerve fibres) 116 positive diffuse – Ob. 20 X, (H) detail of EMA (epithelial membrane antigen) positive diffuse – Ob. 20 X, (I) LEU 7 – zonal positive - Ob. 20 X, (J) CEA (carcinoembryonic antigen) negative - Ob. 40 X

Discussion

Chordomas involving the sellar region are extremely rare, more frequent in younger patients - second to fifth decades of life, with a male/female ratio of 6: 5 (5, 7) Most chordomas involving the sellar region depending on anatomical location and clinical features could be divided in three groups (12): parasellar or suprasellar chordomas with optic tract compression and oculomotor nerve palsy; largely or entirely sellar chordomas – are very rare, mimicking pituitary adenoma with chiasm

compression and hypopituitarism, clival chordomas with brainstem compression and bilateral sixth cranial nerve paresis. Several studies according to Thoudou et al. (12) has pointed the misdiagnosed preoperatively as pituitary adenoma, craniopharyngioma based on clinical symptoms (visual deficit as the most common symptom, hyperprolactinemia or hypopituitarism more commonly observed in the sellar and suprasellar chordomas) and radiologic findings (bone destruction involving pituitary fossa, petrous bone,

orbit, foramen ovale, jugular foramen and even foramen magnum; calcifications in the tumoral mass or sequestered bone fragment) The skull base chordomas tend to present osseous permeation, and have a high rate of recurrence (4-7, 10, 14, 15). Their metastatic potential cannot be estimated by histological examination, except in cases of sarcomatous transformation (1-3, 6, 8, 10).

The imaging method of choice are MRI, also CT – scan with contrast and thin slices for reconstructions (7). A sellar chordoma CT scan may reveal a contrast-enhancing intrasellar mass with bony erosions and calcifications well demonstrated in bone window images (11, 15).

In general surgery should be complete, the capsular tumoral adhesions to chiasma or another nervous structure made this purpose sometimes impossible and postoperative irradiation are recommended despite inconclusive data (7, 10, 12).

The immunohistologic profile of chordoma cells is reactivity to cytokeratins, vimentin, and epithelial membrane (1-3, 7, 9). The only known prognostic study published in 1993 estimated overall survival rates of 51% and 35% at 10 and 20 years, respectively (6). Differential diagnosis should be made with other large sellar and suprasellar tumor mass as pituitary adenomas, especially if a calcified intrasellar tumor with bone erosion is diagnosed; chondroid chordoma, chondrosarcoma with mucoid intercellular substance containing calcifications (5, 7, 14).

Conclusion

Chordomas are midline slow-growing tumors that arise from proximal and distal extreme remnants of the notochord, are locally invasive, may metastasize and are characterized by local recurrence. These tumor can mimic a pituitary adenomas, especially if a calcified intrasellar tumor

with bone erosion is diagnosed. A gross-total resection followed by radiotherapy, has been the accepted treatment for sellar chordomas.

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