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Journal of Cancer Research and Cellular Therapeutics

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Case Report

Intrasellar Chordomas Mimicking Pituitary Adenoma

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Received Date: February 15, 2024; Accepted Date: February 21, 2024; Published Date: February 28, 2024

Citation: A. Lahouaoui, I. Boujguenna, C. Dabajj, A. Fakhri, H. Rais, et al, (2025), Intrasellar Chordomas Mimicking Pituitary Adenoma, *J. Cancer Research and Cellular Therapeutics*. 9(1); **DOI:10.31579/2640-1053/231**

Abstract

Chordomas involving the sellar region are uncommon and has a poor prognosis. In view of its rarity and nonspecific symptoms, clinicians may misdiagnose intrasellar chordoma as pituitary adenoma based on preoperative radiographic images. In this report we present two cases of intrasellar chordomas mimicking nonfunctioning pituitary adenoma.

Keywords: chordoma; sellar tumor; pituitary adenoma

Introduction

Chordoma is a rare bone cancer that is aggressive, locally invasive, and has a poor prognosis. Chordomas are thought to arise from transformed remnants of notochord and have a predilection for the axial skeleton, with the most common sites being the sacrum, skull base, and spine. They represent 1% of all malignant bone tumors and 0.1 to 0.2% of intracranial neoplasms. The incidence of endocranial chordomas presents a male/female ratio of 6: 5, and. The incidence is higher in younger patients (second to fifth decades of life).

Chordomas involving the sellar region are rare, clinically, they may resemble patients with pituitary adenoma.

In this article, we report two cases of intrasellar chordoma simulating anon-functioning pituitary adenoma.

Methods

For microscopic examination, 4 to 6–m sections of formalin-fixed, paraffin-embedded tissue samples were stained with hematoxylin and eosin. For immunocytochemical analysis the immunoperoxidase study (automated technique: OMNIS Agilent station) was used.

Case Presentation

Case 1

A 42-year-old woman presented with hemiparesis and left ophthalmoplegia. She underwent magnetic resonance imaging which revealed a sellar and supra sellar lesional process measuring 48x37x34mm. It infiltrated the sphenoid bone, filled the optochiasmatic cistern superiorly, compressed and displaced the optic chiasm, laterally invaded the cavernous cavities with a mass effect on the left temporal lobe, and posteriorly filled the pre-pontine cistern with compression of brainstem in relation to a pituitary macroadenoma (Fig.). No endocrinopathy was noted and serum pituitary hormone levels were normal. Initially, the lesion was considered to be a non-functioning pituitary adenoma. The patient underwent surgery. The initial anatomopathological result concluded that the morphological appearance was consistent with a pituitary adenoma. 12 months after the initial surgery, the patient's symptoms worsened. A follow-up MRI showed an increase in the size of the locally advanced sellar and supra sellar process, with post-therapeutic changes favoring recurrence with suspected malignant degeneration. An external review with immunohistochemistry was requested. Microscopic examination revealed a tumour proliferation arranged in lobules and short cords. The lobules were delimited by fine fibrous bands containing a few regular vascular structures. The tumour cells are epithelioid in appearance, with round oval anisokaryotic nuclei that are hyperchromatic in places and have vesicular chromatin that is discretely nucleated elsewhere. The mitotic index is estimated at less than 2 mitoses/10 fields. The cytoplasm is abundantly eosinophilic and locally clarified. In addition to these predominant cells, there are phisaliform cells with round pycnotic nuclei and multi-vacuolar cytoplasm. The stroma is hyalinised fibro myxoid in places. There is no necrosis or fusiform component (Figure.). Special alcian blue coloration revealed large foci of extracellular mucins (Figure.). The immunohistochemical study showed diffuse positivity of the tumour cells for anti-cytokeratin, anti-EMA, anti-S-100 protein and anti-vimentin antibodies (Figure.). A diagnosis of conventional chordoma was made.

Case 2

This 28-year-old woman was admitted with intracranial hyper tension syndrome. MRI revealed an intrasellar sellar and supra sellar mass invading the cavernous sinuses and protruding into the sphenoid sinus, measuring 2 cm in diameter. Endocrine evaluation showed normal findings. Pathological findings. Microscopically, the tumour was composed of lobules of large cells with uniform nuclei mainly embedded in a myxoid stroma mixed with vacuolated cells (physaliphores). In

addition, the lobules of the tumour cells were sometimes separated by bundles of fibrous tissue. The tumour cells contained abundant glycogen. Immunostains were positive for EMA, S-100 protein and pancytokeratin. A diagnosis of conventional chordoma was made.

Discussion

Chordomas are rare bone tumours that develop on the remnants of the embryonic notochord. They are slow-growing tumours, invading neighbouring structures and rarely metastasizing to distant sites. However, they present a high risk of local and locoregional recurrence after treatment. They are often found in the skull base (35%-40%) and dorsal sacrum (40%-50%). Chordomas involving the sellar region are extremely rare. According to a recent review, only 22 patients have been described as having a predominantly intrasellar chordoma since 1960, and 43% of patients were primarily diagnosed with a pituitary adenoma. This rate of misdiagnosis could arise mainly because intrasellar chordomas mimic the clinical and imaging presentations of pituitary adenomas, as described in the two present cases. Chordoma can be seen at any age, with a median age of diagnosis of 60. Men are clearly more affected than women. Our case involved two women, aged 42 and 28. Symptoms depend on the location of the tumour. Magnetic resonance imaging (MRI) is the main diagnostic method, but anatomopathological examination remains the fundamental and indispensable element for the diagnosis of chordomas, as indeed for all bone tumours. According to the WHO, "chordomas are malignant tumors with notochordal differentiation". Macroscopically, they are generally lobulated tumours with a soft, gelatinous consistency, greyish or bluish-white in color. Microscopically, the tumor cells are large, with well-defined cytoplasm, homogeneous eosinophilic or clarified, containing one or more optically empty vacuoles pushing back the nucleus, sometimes distorting the cytoplasm to give the appearance of a physaliphore cell. These cells are grouped in cohesive clusters of variable size, or form single-cell trabeculae, or isolated cells, on a myxoid matrix of highly variable abundance from one tumor lobule to another within the same tumor.

Nuclei are variable in size and contour, somewhat irregular and hyperchromatic. Mitosis, hemorrhagic or necrotic inflammatory changes, sometimes extensive, and apoptosis may be observed. In chordomas, pancytokeratins and cytokeratin 19, EMA (epithelial-membrane antigen) and vimentin are almost constantly expressed, but sometimes only focally. Brachyuria is a more recent marker, highly specific for chordoma, with a sensitivity of around 90.2%. Treatment of sellar chordomas consists of transsphenoidal surgical removal followed by radiotherapy. The prognosis is generally poor, given the importance of the cerebral structures likely to be compressed by the tumor, and also because total excision is practically impossible.

Chordomas, especially their cranial forms, are uncommon tumours. Depending on their topography and radiation, they may simulate a pituitary adenoma, and should be included in the differential diagnosis of tumors of the sellar region.

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DOI: 10.31579/2640-1053/231