A DESTRUCTIVE CENTRAL SKULL BASE PROCESS IN A YOUNG PATIENT WITH SYSTEMIC ENDOCHONDROMATOSIS

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Abstract

Introduction: This is a case of a 20-year-old female with systemic endochondromatosis (Ollier’s syndrome) who presented with a central skull base lesion.

Methods: Review of patient’s clinical, radiographic, and histopathologic records as well as literature review.

Results: The patient underwent evaluation for seizures, galactorrhea, and headaches. Imaging revealed a destructive central skull base lesion for which the patient underwent endonasal endoscopic partial resection. An unusual fullness in the adjacent adenoid areas was also resected. Final pathology revealed skull base enchondroma and benign nasopharyngeal tissue. The patient improved symptomatically after surgery.

Conclusion: Patients with systemic endochondromatosis can develop similar lesions at the skull base, and management includes removal for biopsy.

Radiographic Images

The computed tomography (CT) images in the axial (A), sagittal (B), and coronal planes (C,D) reveal a large, expansive, destructive mass involving the central skull base, including the clivus, pituitary fossa, paracalival region (arrows). There was also distinct soft-tissue fullness in nasopharynx (asterisk).

Introduction

Central skull base lesions can be challenging diagnostically and therapeutically. Differential diagnosis includes chondrosarcoma, plasmacytoma, intracranial lymphoma, invasive pituitary macroadenoma, nasopharyngeal carcinoma, etc. Work-up includes full history, physical exam, appropriate imaging, and possible endoscopy.

This patient had a lifelong history of systemic endochondromatosis. Chondromas are tumors of hyaline cartilage that most commonly arise in the hand phalanges, and are classified as enchondromas when they originate from the medulary canal. Ollier’s disease, first described in 1898, commonly presents in the first decade of life and is defined as 3 or more enchondromas in asymmetric distribution, though its presence can be quite variable. Maffucci syndrome is enchondromatosis with soft tissue hemangiomas.

Clinical History and Physical Examination

The patient is a 20-year-old female with an expansive and destructive lesion of the central skull base. The patient has a lifelong history of Ollier’s syndrome, manifested by multiple and progressive deformities of the left hand and leg, which have required approximately 10 surgeries over her lifetime. She has experienced associated problems beyond the bony lesions, including Maffucci syndrome-related changes of the left hand, leukopenia, scoliosis, and the fact that her legs are of very disparate lengths.

She was in her usual state of health when she developed headaches that she refers to as migraines, and then had a grand mal seizure. Symptomatic complaints included visual blurring, galactorrhea, headache, seizures, partial upper airway obstruction and snoring. Seizures were managed well medically and the patient was referred by neurosurgery colleagues regarding possible endoscopic biopsy after initial imaging was abnormal, revealing a lesion in the clivus extending into the suprasellar area.

She also had a nasopharyngeal lesion that appears distinct from the skull base mass but could be related. This was causing restricted nasal airway manifested by snoring and probably obstructive sleep apnea on clinical grounds.

One of the challenging problems in this particular patient’s case is that she is extremely petite, standing only about 4 feet 8 inches tall, as part of her Ollier syndrome. In this case, her nasal development was incomplete and the nasal airway was extremely narrow on both sides, making an endoscopic approach significantly more difficult than usual.

She is a very pleasant, alert, and oriented African-American female of very short stature (4 ft, 6 inches) and has deformity of left hand and leg. The left hand and leg are enlarged, deformed with expansile changes consistent with her known diagnosis of Ollier’s syndrome.

External nose appeared normal.

Endoscopic examination of the nose in clinic revealed the right nasal cavity is stenotic and it was very difficult to pass the small endoscope. The left nasal cavity was compromised by a deviated septal spur as well. In the nasopharynx, there was a discrete mass slightly more left-sided than right, too asymmetric to be adeno.

Operative Technique & Pathology Findings

The patient underwent transnasal endoscopic surgery with extensive tissue sampling of the bony lesion as well as nasopharyngeal mass. The clival mass was diagnosed as a very low grade cartilaginous lesion, most consistent with enchondroma described as mature cartilaginous neoplasm composed of chondrocytes with variable cellularity and an abundant hyaline cartilaginous matrix.

Pathology of the nasopharyngeal mass returned as benign lymphoid tissue with follicular hyperplasia with sections remarkable for a hyperplastic appearing lymphoid tissue. Work-up for lymphoma was negative.

Outcome and Discussion

Chondromas are tumors of hyaline cartilage that most commonly arise in the hand phalanges, and are classified as enchondromas when they originate from the medulary canal. The pathophysiology is not clear, but the end result is that long bones expand in diameter but tend not to lengthen.

In this patient, the clival bone underwent expansive changes with pituitary changes and the patient suffered from seizures and headaches. Fortunately, these have resolved since near-total excision of the lesion.

The association of enchondromas and chondrosarcomas has been described. Chondrosarcomas of the nasal cavity and intracranial cavity have been described associated with Ollier disease and Maffucci syndrome. Ranger and Szymczak, in comparing intracranial neoplasms of Ollier disease and Maffucci syndrome, noted that malignancy was significantly more common in the latter group. Fortunately, this patient had benign histopathology.

Her post-operative course over the last 12 months has been uneventful. She is being followed with serial endoscopic and radiographic examinations.

This clinical entity of skull base enchondroma with local destruction is rare, and it can be addressed appropriately with comprehensive clinical evaluation including endoscopy, radiographic studies, and surgical biopsy.

References