Clinical Challenge | RADIOLOGY

A Young Man With Proptosis Causing Decreased Visual Acuity

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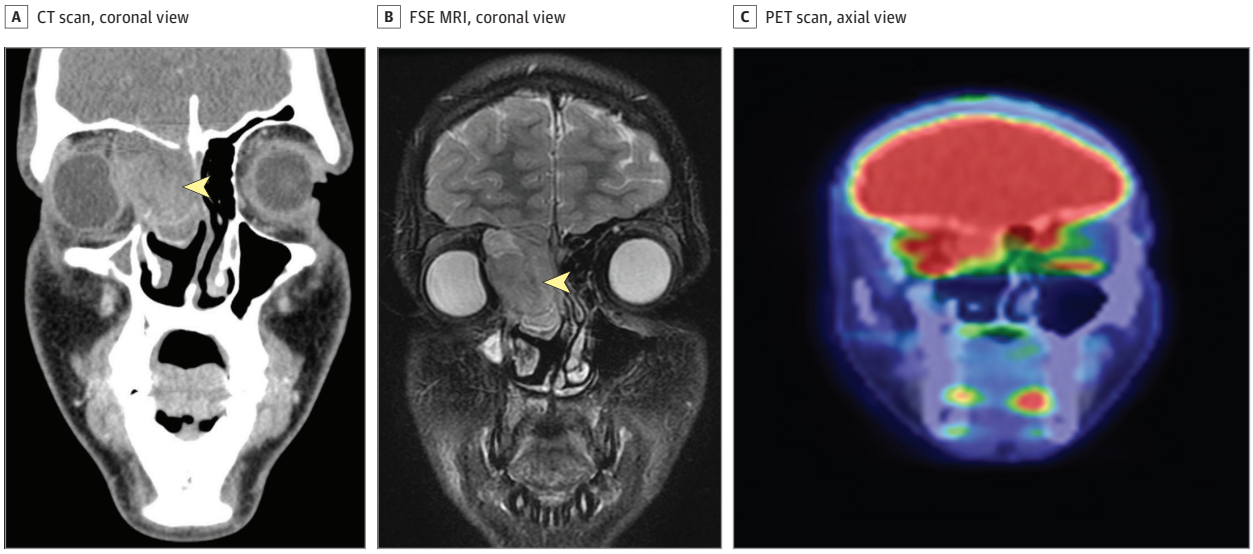

Figure. A, Postintravenous contrast computed tomographic (CT) scan of the face showing a relatively homogenous mass of the right ethmoid complex with invasion of the orbit and extension to the nasal cavity (arrowhead). B, Fast spin echo (FSE) T2 magnetic resonance image (MRI) of the right ethmoidal/orbital soft-tissue mass (arrowhead). C, Positron emission tomographic (PET) scan of the face with fludeoxyglucose F 18 showing increased uptake within the right ethmoid air cells and nasal cavity, and in the medial right orbit.

A teenage boy was referred with a progressive 2-week history of blurred vision in his right eye and right ear pain, which had worsened in the past 7 days. This change prompted a visit to an optometrist, who found a mild decrease in visual acuity and minor cataract in the right eye. His primary care physician diagnosed his ear pain as acute otitis media and prescribed a course of amoxicillin-clavulanate; however, the young man's vision continued to deteriorate and became associated with the onset of right eye swelling and forehead tenderness. A repeat eye examination by an ophthalmologist was performed and showed normal peripheral visual acuity with a decrease in central vision and confirmed proptosis. The patient had not experienced nasal congestion, epistaxis, weight loss, fevers, or chills. Physical examination included right eye proptosis and restricted eye movement. A computed tomographic (CT) scan with contrast of the face showed a small relatively homogenous mass in the right anterior nasal cavity invading the right orbit, and a magnetic resonance image (MRI) with contrast showed a right superior nasal cavity mass, with invasion of the right orbit and right ethmoidal sinus with extension to the overlying soft-tissue scalp (**Figure**). A 30° endoscope was used to visualize the superior nasal cavity and revealed a fleshy red soft-tissue mass. Several biopsy specimens were taken and sent for frozen pathologic evaluation.

WHAT IS YOUR DIAGNOSIS?

A. Esthesioneuroblastoma

B. Rhabdomyosarcoma

C. Ethmoid mucocele

D. Sinonasal undifferentiated carcinoma

Diagnosis

B. Rhabdomyosarcoma

Discussion

Rhabdomyosarcoma (RMS) is the most common pediatric soft-tissue sarcoma and can occur anywhere in the body. It is a tumor of mesenchymal origin and develops in the striated muscle.[1] The 2 most common locations are the genitourinary system and the head and neck (typically spreading to parameningeal spaces and meningeal areas).[2] There are 4 histological subtypes of RMS: embryonal, alveolar, spindle cell or sclerosing, and the pleomorphic tumor.

Pediatric sinonasal tumors remain difficult to diagnose. Rhabdomyosarcoma usually mimics common allergic symptoms of nasal congestion; however, 53% of patients may present with more serious neuropathic abnormalities.[1,3] Reilly et al[4] looked at nonorbital tumors of the head and neck and found overall 5-year survival rate of approximately 75%.

To diagnose RMS, the clinician must identify and localize the tumors. The best imaging modalities that are currently available are CT and MRI. Gomaa et al[5] showed that these imaging modalities are needed to determine the location and the extent of metastases of sinonasal tumors.

Magnetic resonance imaging has greater sensitivity than CT to distinguish inflammatory tissue from tumor and to demonstrate intracranial extension as well as a better ability to determine involvement of the neural and vasculature structures of the head and neck region.[5,6] Both CT and MRI can be used to determine the extent of involvement of the surrounding tissues and the aggressiveness of the tumor. For example, while bony remodeling indicates a slow benign process, as may occur in mucocele, bony destruction usually indicates a more malignant process.[6]

Certain imaging characteristics, in addition to the patient history and presentation, may be indicative of the type of tumor present. Esthesioneuroblastoma is a tumor of neural crest origin. The presence of a mass in the caudal nasal cavity with extension to the neurocranium is highly suggestive of esthesioneuroblatoma.[7] Sinonasal undifferentiated carcinoma (SNUC) is a tumor of paranasal sinuses. It has radiographic features of a soft-tissue mass with heterogenous enhancement on CT. On MRI, SNUC appears isointense to muscle on T1, likely hyperintense on T2, and enhances heterogeneously. Mucoceles have cystic characteristics on both CT and MRI. If not infected, mucoceles are hypodense on CT and show increased T2 signal on MRI. They do not enhance on either CT or MRI. Chronically, mucoceles become hyperdense on CT and hypointense on T2 MRI. Rhabdomyosarcomas, as in the case described herein, are usually seen as a solitary soft-tissue mass. Although perineural, adjacent osseous destruction or metastatic spread can be visualized, imaging characteristics are nonspecific.

In this case, the contrast-enhanced CT, as seen in the Figure, A, showed a mass within the right ethmoid complex/nasal cavity with invasion of the right orbit and right frontal sinus. There was invasion of the right frontal scalp (not shown) through the outer cortex of the frontal sinus. The MRI image in the Figure, B, did not show cystic or necrotic components within the mass.

Positron emission tomography (PET) (Figure, C) performed after tissue biopsy but before final diagnosis showed increased glucose uptake within the mass, which is highly concerning for a malignant process. Multiple neck nodes and a superficial left femoral node also showed increased glucose uptake, causing concern for metastasis.

There are inherent limitations to imaging because the diagnosis of RMS will always need to be made based on a biopsy specimen of the tumor. However, the CT, MRI, and PET scan helped ensure that the biopsy could be safely performed without the risk of bleeding, as would occur with a vascular tumor, and minimized risk of cerebrospinal fluid leak, which could occur with dural invasion from any malignant sinonasal mass.

ARTICLE INFORMATION

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