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Intracranial Complications from Temporal Bone Cystic Angiomatosis

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Case Report

A 47-year-old male presented to a local hospital with a one-year history of progressively worsening right-sided headaches and more recent onset of gait instability along with intermittent lower extremity weakness. Computed tomography (CT) of the head was obtained which showed an expansile mass ($3.9 \times 7.5 \times 4.7$ cm) centered in the mastoid portion of the temporal bone with involvement of the anterior portion of the occipital bone. (Figure 1) The patient was then transferred to our institution. An MRI was obtained which revealed a T2-weighted hyperintense mass that avidly enhanced on T1-weighted images with gadolinium (Figure 2A, B, C). There was severe mass effect of the right cerebellum causing compression of the fourth ventricle and cerebral aqueduct with resultant hydrocephalus. Subsequently, the patient underwent placement of external ventricular drain. The patient did not complain of hearing loss and there appeared to be no involvement of cranial nerve VII or VIII on imaging. Additional CT imaging of the thoracic and abdominal cavities found multifocal mixed lytic and sclerotic osseous lesions of vertebral bodies, ribs, and bony pelvis. CT-guided biopsy of the left iliac wing lesion showed findings consistent with hemangioma. The decision was made to remove the right skull base lesion to obtain a definitive diagnosis and to relieve the compressive effects of the mass. Prior to surgery, neuro-interventional angiography was performed showing the lesion to be hypervascular and supplied by the right occipital artery and muscular branches from the right vertebral artery (Figure 2D). The lesion was then embolized with Onyx (Covidien, Plymouth, NH).

The patient underwent a right sided craniectomy of the occipital and temporal bones with gross removal of the tumor to the level of the dura. The bony removal continued to the transverse and sigmoid sinuses with further bony removal from anterior to sigmoid into the mastoid cavity. This allowed the posterior fossa to return to its normal position. Minor dural defects were closed primarily and there was no cerebrospinal fluid (CSF) leak at the end of the procedure. Adipose tissue was placed into the defect and secured with a titanium mesh

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plate. Post-operatively the patient had a grade 1/6 House-Brackman facial nerve function and had the external ventricular drain removed on post-operative day 4. The patient was then discharged home. However, two days after discharge the patient presented back to the ED with a CSF fistula presenting through the postauricular incision. The patient was found to have continued hydrocephalus that was presumptively due a persistent imbalance in production and reabsorption. The patient then underwent placement of a ventriculoperitoneal shunt. Since placement of the shunt, the patient has subsequently done well with minimal sequelae from surgery. The final pathology of the excised portion of the temporal bone were consistent with cystic angiomas (CA).

Discussion

This represents the first case report of CA causing posterior fossa and cerebellar compression leading to hydrocephalus. Cystic angiomas, sometimes referred to as cystic lymphangiomas, is a rare disease characterized by disseminated angiomatous or lymphangiomas cysts that affect the skeleton - especially the long bones, pelvis, vertebrae, and skull. (1) This process typically presents in the first decade of life, although many cases occur later in life indicating a possible congenital and acquired etiology, respectively. There are published case reports on CA featuring CT imaging and radiographs of skull lesions, but no head and neck magnetic resonance imaging (MRI) imaging of these lesions have been published. On MRI, CA lesions are typically T2 hyperintense and enhance after gadolinium on T1 weighted images. (2) CT may show single or multiple lytic lesions with either peripheral thinning or sclerosis. These radiologic findings are quite unique and combined with findings of systemic lesions, may lead to a diagnosis. Obtaining an adequate specimen for histologic diagnosis may be quite difficult in this condition with multiple attempts being commonly reported. (3)

The differential diagnosis for this lesion on temporal bone imaging includes chordoma, chondromyxoid fibroma, hemangiopericytoma, intraosseous meningioma, histiocytosis X, hyperparathyroidism, polyostotic fibrous dysplasia, metastases, and lymphomas of bone. Clinical symptoms can vary based on the location of lesions and include pain, weakness, swelling, difficulty walking, and scoliosis. Patients are usually diagnosed when lesions are discovered incidentally or when they lead to pathologic fractures. CA treatment is supportive and targeted at controlling symptoms, such as fixation of fractures or excision of soft tissue lesions. Bisphosphonates, radiation, and interferon have been reported treatments as well as monitoring the patient for spontaneous arrest of lesions. (4). In conclusion, CA should be included in a differential diagnosis of lytic lesions of the skull base.

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FIG 1.

Computed tomography of right temporal bone A, Axial view of skull showing R temporal bone lesion with mixed osteolysis and sclerosis of the bone and peripheral thinning. There is also involvement of the right anterior occipital bone. B, Axial view of lesion magnified to show the moth-eaten appearance of the temporal bone. C, Coronal view

FIG 2.

Magnetic resonance imaging of right temporal bone A, T2 weighted axial view demonstrating a hyperintense lesion after contrast administration. B, T1 weighted coronal view. C, T1 weighted coronal view showing contralateral temporal bone and cervical vertebrae lesion (arrow). D, Angiography of lesion demonstrating increased vascularity diffusely.