

Oncogenic Osteoblastoma: A Rare Clinical Entity in the PNS Arising from Ethmoidal Sinus

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ABSTRACT

Oncogenic osteoblastoma is a rare clinical entity in the peripheral nervous system (PNS). Its presentation is like a nasal polyp. One should keep the diagnosis of oncogenic osteoblastoma in mind as a differential diagnosis for mass in nasal cavity. Excessive bleeding during surgery should arouse the suspicion and the pathologist has to be sounded. A preliminary biopsy of the mass is to be considered. Immunohistochemistry (IHC) should be done, if the suspicion is strong.

Keywords: Oncogenic osteoblastoma, IHC, PNS, Nasal polyposis, Nasal mass.

CASE REPORT

Herein we present a case report of a rare entity in the paranasal sinus. A 30-year-old female patient came with history of nasal obstruction since 2 months. She also had occasional nasal bleed, which was very scanty and insignificant. Loss of sensation of smell, sneezing and nasal discharge were part of her daily routine along with slight nasal intonation of voice. There were no significant aural complaints and no contributions were obtained regarding symptomatology of throat.

Systemically, she came with severe bone pains all over the body, especially in the pelvic region. In view of the pain she could not climb stairs and had a waddling sort of gait. Patient gave a past history of admission to a premier tertiary care hospital for evaluation of lower backache, which had occurred after trauma. She was investigated and found to be anemic.

Investigations done were Hb –7.9 g/dl, Ca –8.8 mg/dl, iPTH 27.7 pg/ml, SpO₄ –1.6 mg/dl, ALP –108 IU, blood urea –22 mg/dl, serum creatinine –0.6 mg/dl, urinary calcium 67 mg/24 hours, urinary pO₄ – 449 mg/24 hours.

X-ray: Right leg upper segment revealed lytic lesion, thoracolumbar spine—D11, D8 endplate changes were noted.

Bone scan: Multiple hot spots were seen in pelvic, scapular, spinal, iliac bone, fibula and upper-third of ribs (Fig. 1).

MRI: Diffuse and right lateral disk bulge at L4-L5 with mild impression on thecal sac and right foraminal hollowing. Diffuse disk bulge at L5-S1 with mild impression on thecal sac.

All these aspects gave a probable differential diagnosis of metabolic bone tumor/Koch's/multiple skeletal metastasis. Patient was treated with calcium and vitamin D supplements, improved symptomatically and was discharged.

A principle diagnosis of osteomalacia⁵⁻⁷ was done with no neurological deficits. Further evaluation revealed normal functioning thyroid. Sputum for AFB did not yield any positivity.

A Mantoux test yielded a positive report with 25 mm induration at the site of test.

However, much ado was not done regarding this report.

At this stage physician opinion was taken to rule out multiple myeloma.

Bone marrow study was normal and X-ray of skull was also not contributory. As part of screening procedure, USG of abdomen/pelvis revealed a bulky uterus not contributing to a diagnosis. Hemogram was normal except for a high ESR and anemia. Total proteins/protein fractions were normal which were assessed by serum protein electrophoresis. Repeat serum calcium levels were low normal.

A CT scan of PNS was done which gave a provisional diagnosis of juvenile nasopharyngeal angiofibroma (JNA).

Clinical examination of mass: Pale pinkish mass was filling both the nasal cavities with tinges of ooze on probing and pain due to the pressure effect, not able to get all around the swelling. Too much probing was not done for fear of inducing bleeding and thought shifted to diagnosis of mass being JNA.

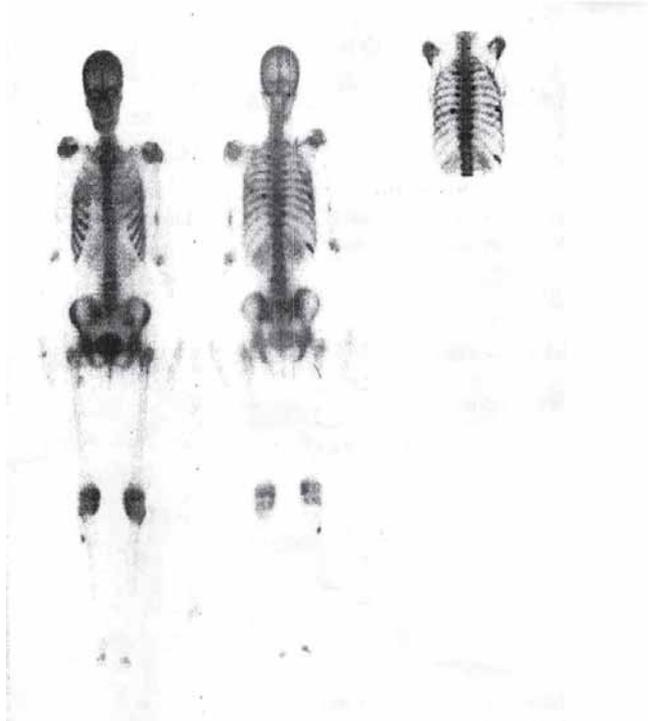


Fig. 1: Pet sensation (Courtesy: Center for molecular imaging)

After CT scan, the patient was referred to NIMHANS for embolization of feeding vessel of suspected angiofibroma. Selective right ICA, ECA angiogram was done. There was no evidence of tumor blush.

A differential diagnosis of right sinonasal mass was arrived at.

After the evaluation for anesthesia and physician's fitness, patient was shifted for endoscopic clearance of nasal mass. Intraoperatively, there was brisk bleeding and consistency of mass was near rubbery. Entire areas of nasal cavity per se, maxillary antri, ethmoids and sphenoid sinuses were cleared. After clearance of the entire tumor mass, bleeding became negligible. There were empty spaces appear with expanded nasal cavity postsurgically. Removed specimen was sent for HPE and a probable differential diagnosis of rhinosporidiosis was also suggested. Preliminary report from Department of Pathology was inconclusive. The slides and blocks were sent for revision HPE/marker/IHC and the diagnoses of chondroblastoma with aneurysmal bone cyst-like areas, probably arising from ethmoid bone was made.

Microscopic evaluation showed clear cell with numerous ectatic blood vessels.

IHC was done. Neoplastic cells express vimentin and were negative for S-100, CK and EMA.

Tumor⁴ cells were negative for markers of epithelial differentiation (CK, EMA), myoepithelial differentiation (P63, High mol wt, cytokeratin, calposin, S-100 protein) and myogenic marker (Desmin, SMA).

OBSERVATION

After complete removal of nasal mass, patient was asymptomatic, nasal obstruction was relieved totally and the most striking part was the disappearance of bone/joint pains. The waddling gait also disappeared and patient was walking without difficulty. Follow-up period of 2 years, patient has no recurrence of mass or joint pain.

DISCUSSION

Phosphaturic mesenchymal tumors (mixed connective tissue) PMTMCT are extremely rare tumors although their exact incidence is difficult to determine. Most cases occur in middle-aged adults with small female preponderance; 5% occur in craniofacial sinus. They usually present as small, in apparent lesion that require very careful clinical examination and radionuclide scan for localization in some cases.^{1,2}

Long history of osteomalacia is present in almost all cases. Histologically, they are tumors without hemangiopericytoma, like vessels or osteoclasts and cytologically banal neoplasms with possible worrisome aspects (e.g. osteoid like matrix) and frankly sometimes sarcomatous tumors. FGF 23 gene and protein expression provide additional support.³

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