CASE REPORT

Fibrous Dysplasia Involving Eustachian Tube on ^{99m}TC-MDP Whole Bone Scintigraphy and SPECT/CT: A Rare Case of Tinnitus

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ABSTRACT

Tinnitus is a phantom auditory perception in the absence of external stimuli which can be caused by various etiologies including infectious causes, neurological etiologies such as whiplash injury and acoustic neuroma, side effects of certain medication and mechanical causes. Neoplasms that block the Eustachian tube (ET) may also lead to tinnitus. This is a case of fibrous dysplasia (FD) seen on Technetium-99m methyl diphosphonate (^{99m}Tc-MDP) whole body bone scintigraphy and SPECT/CT causing blockage of the ET. Fibrous dysplasia (FD) is a rare, congenital, and benign neoplasm that affects the bones. FD can be broadly categorized as mono-ostotic and poly-ostotic types. It often extends from its primary site into other bones and may cause symptoms due to compression, which may not be seen by conventional radiological imaging. We present a case of FD in a young woman, who had unilateral right-sided tinnitus and headache. Conventional imaging identified a lesion involving the paranasal bones with no involvement of the temporal bones or middle ear. ^{99m}Tc-MDP whole-body bone scintigraphy was performed to delineate the extent of the disease, which revealed complete blockage of the right sided paranasal sinuses that could have led to a Patulous ET (PET), hence indirectly causing tinnitus. This illustrates the role of nuclear medicine imaging, over conventional imaging, in accurately delineating the functional extent of fibrous dysplasia.
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Keywords: Tinnitus, Fibrous dysplasia, Eustachian tube, Pirate sign, Single-photon emission computed tomography

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INTRODUCTION

Tinnitus is a phantom auditory perception in the absence of external stimuli which can be caused by various etiologies including infectious causes, neurological etiologies such as whiplash injury and acoustic neuroma, side effects of certain medication and mechanical causes. Neoplasms that block the Eustachian tube (ET) may also lead to tinnitus (1). One rare example of neoplasm that can block ET is fibrous dysplasia (FD). FD can be diagnosed using radiography, whereby the radiologic characteristics can be diverse. Typically, the appearance is of an expansile lesion arising from the medulla and progresses to the cortex, which has a cystic appearance surrounded by cortical thinning. In the event of a previous fracture, the cortex can appear thickened. FD typically demonstrates a 'ground-glass appearance' and may have variable internal calcifications on radiographs or computer tomography (2). Nevertheless, it is useful to perform tagged Technetium-99m radionuclide whole-body bone scintigraphy (WBS) to illustrate the extent of the disease. There is a pathognomonic feature of FD in the craniofacial bones which gives the appearance of 'pirate sign', which is useful to be recognized. (3)

CASE REPORT

A previously well 25-year-old female presented with progressive right-sided tinnitus that had been ongoing for several months. It was associated with right-sided nasal blockage and worsening generalised headache. There was neither a history of epistaxis nor history of constitutional symptoms. She sought ENT treatment at a nearby hospital. On examination, there was a prominent swelling over the right maxillary region extending over the right orbital region. There were no overlying skin changes and the swelling was non-tender upon palpation. Ophthalmology examination revealed intact visual fields and visual acuity. There was no neurological deficit detected. Rinne and Weber's tests were negative. Her routine blood investigations were within normal limits. Patient denied any history of recent trauma. She also did not have any family history of similar bone

condition or malignancy.

Subsequently, she was referred for an MRI brain, which detected an expansile bony mass having 'ground glass appearance' arising from the right inferior turbinate, right middle turbinate, ethmoidal sinus and extending to the anterior cranial fossa. At that time a provisional diagnosis of FD of the right inferior turbinate was made. Soon after, she was scheduled for an examination under anaesthesia and a biopsy of the mass was taken. Intraoperative findings showed a right nasal cavity mass with enlarged and hypertrophied inferior and middle turbinates. The inferior turbinate appeared bony and thickened. HPE of the biopsied specimen revealed a moderately cellular fibrous stroma surrounding irregular, curvilinear trabeculae of woven bone, hence confirming the diagnosis of FD.

She was then referred to us for a ^{99m}Tc-MDP whole-body bone scintigraphy to look for the extent of the lesion and to identify other potential sites that may be affected by the condition. After intravenous injection of 18 mCi of ^{99m}Tc-MDP, the bone scintigraphy was performed after 3 hours of uptake time, in the anterior and posterior planar views. The bone scintigraphy revealed a large solitary, focus of increased tracer uptake seen at the right side of the paranasal sinuses, the right orbital roof, and the medial aspect of the right maxillary region, giving the appearance of the 'pirate sign' (Fig. 1). Faint tracer uptake was also noted extending posteriorly along the right sphenoid bone. A complementary single-photon emission computed tomography / computerized tomography (SPECT/CT) of the skull was performed for anatomical localization to evaluate the extent of the skull bone involvement. SPECT/CT revealed tracer uptake in the expansile lesion with 'ground-glass appearance' arising from within the completely obstructed right sphenoid sinus (Fig. 2). Tracer uptake was also noted over the right superior maxillary ridge, the medial wall of the right maxillary sinus, and the right supraorbital ridge extending posteriorly to the right sphenoid bone, with dilated and patulous ET (Fig. 2).

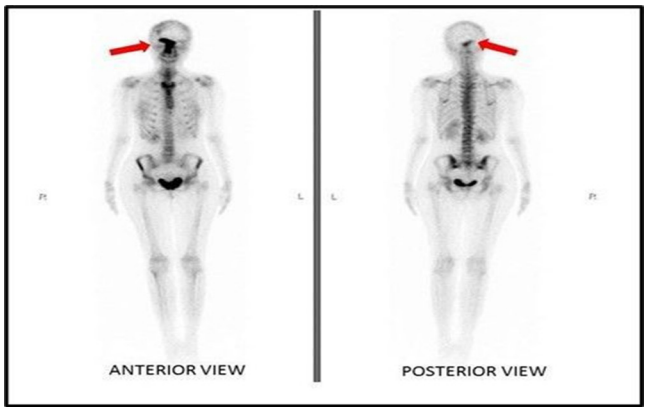

Fig. 1: Planar bone scintigraphy in anterior and posterior views show an intense hot spot (red arrow) demonstrated at the right supraorbital, right sphenoid and the right maxillary regions, which appears as a 'pirate sign'.

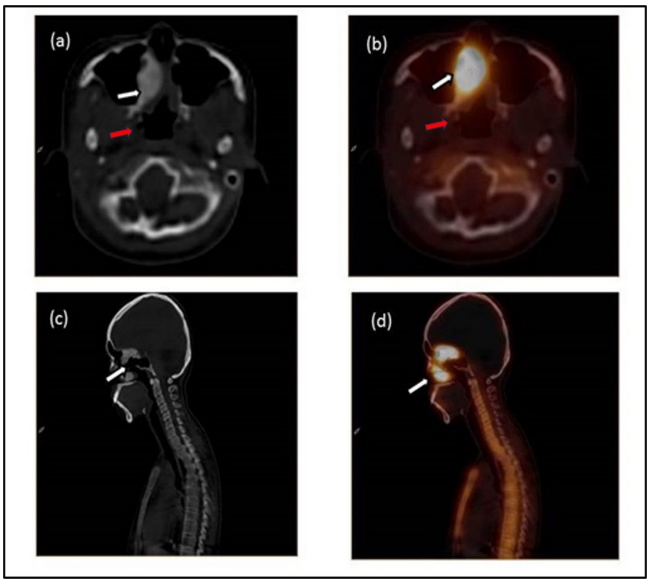

Fig. 2: CT and fused SPECT/CT images of the region of interest at the level of the nasopharynx. (a) Axial CT scan showed an expansile lesion with 'ground-glass appearance' at the right sphenoid sinus (white arrow). There is also evidence of expansion of the right Eustachian tube giving rise to Patulous Eustachian tube (PET) (red arrow). (b) Fused SPECT/CT images in axial view that revealed an intense hot spot at the right sphenoid lesion (white arrow). There is also evidence of expansion of the right Eustachian tube giving rise to Patulous Eustachian tube (PET) (red arrow). (c) Sagittal CT scan showed the extent of the lesion involving the right sphenoid, right maxillary and right supraorbital regions (white arrow). (d) Fused SPECT/CT image in sagittal view identified a metabolically active, large mono-ostotic lesion involving the skull and facial bones (white arrow).

DISCUSSION

Fibrous dysplasia (FD) is a rare, congenital, and benign neoplasm that affects the bones. It is caused by the failure of the process of normal osteoblastic differentiation, leading to focal proliferation of fibrous connective tissue interspersed with islands of immature woven bone. FD can be broadly categorized as mono-ostotic FD and poly-ostotic FD and may also present as part of a condition known as the McCune-Albright syndrome, which is associated with 'café-au-lait' spots and precocious puberty. FD does not have a predilection for any particular bone in the body; however, it commonly affects the skull and facial bones, followed by the long bones, pelvis and ribs. Many bones can be affected simultaneously but the disease itself is usually unilateral and the pattern of involvement does not commonly change with age.

FD is a metabolic aberration that is non-hereditary, having no gender predilection and is typically detected by the third decade of life. Facial and cranial bones involvement occur in the majority of the polyostotic type and in a third of monostotic type of cases (2). In monostotic type, jawbone lesions commonly involve the maxilla comparatively to the mandible. Hence, the progression of FD can slow-down or stop altogether after puberty or bone maturation. However, it has been known for disease to progress into old age. Therefore,

imaging using whole body bone scan plays an important role to delineate the extent of the disease and aid for further decision-making management (2). Whole body bone scans can be useful for assessing the activity of FD because the uptake is reduced in the quiescent state (2). Furthermore, bone scans can help in characterizing the lesion and differentiating FD from other conditions that have similar radiological appearances such as nonossifying fibroma or a fibrocortical defect, whereby these other conditions generally demonstrate minimal or absent uptake (2).

The classical appearance of FD involving the facial bones, particularly the sphenoid wing as seen on Tc-99m bone scintigraphy has commonly been described as the 'pirate sign' (3). Nevertheless, there are other conditions that may have similar bone scintigraphy appearance such as Paget's disease (4). Planar bone scintigraphy is a nuclear medicine scan that enables the detection of abnormal osteoblastic activity. Common indications are to look for bone metastasis in oncology patients, to characterize indeterminate bone lesions and to look for treatment response for bone cancers. Other non-oncology indications include investigation of bone pain of unknown cause, metabolic bone disease and characterization of congenital bone lesions. The role of WBS in this patient was to delineate the extent of the lesion and to look for other potential sites affected by the condition (5).

Typically, cranio-facial FD involves the sphenoid bone, hence the word "pirate signs" that is reminiscent of an eye-patch worn by pirates. However, other facial bones have also been frequently involved such as the temporal bone, external auditory canal (EAC) and middle ear ossicles which ultimately leads to tinnitus (4). The pathophysiology of tinnitus and hearing loss in the previously reported FD cases is due to the direct involvement of the disease into the EAC leading to obstruction and secondary cholesteatoma formation with associated middle ear destruction.

In this particular case, there was no direct involvement of the middle ear. The aetiology of tinnitus is postulated to be due to increased pressure in the nasopharynx caused by paranasal sinuses blockage, leading to imbalance of pressure in the middle ear. Consequently, this increased pressure caused widening of the ET i.e., Patulous Eustachian tube (PET). PET occurs when the ET is permanently open and patients experience a sensation of increased pressure, a form of tinnitus caused by awareness of the blowing sound of their own breathing, and sometimes autophony. Normally, the ET is closed at rest in order to protect the middle ear from nasopharyngeal secretions. The ET opening occurs only briefly during swallowing and Valsalva. In other words, if this protective closure of the ET is disrupted, then patients may experience PET dysfunction (1).

PET has been known to occur in patients with nasopharyngeal carcinoma, leading to symptoms that includes tinnitus and hearing loss as a consequence of middle ear effusion (MEE). MEE are among the most common findings associated with NPC as it causes negative pressure in the tympanic cavity leading to PET. This can be a sequel from direct tumour invasion or compression of the Eustachian tube. To the best of our knowledge, this is the first case report to demonstrate tinnitus due to PET that was indirectly caused by FD in the paranasal sinuses.

In summary, the addition of complementary SPECT/CT helps to enhance the sensitivity for FD lesion detection by improving the anatomical localization of scintigraphy findings based on the added diagnostic information provided by multiplanar reformatted CT. In fact, SPECT/CT can exert a pivotal role in the management of FD including detection of malignant transformation.

CONCLUSION

Planar bone scintigraphy with complementary SPECT-CT is a useful imaging tool for delineating the extent of fibrous dysplasia. Its advantages in detecting the pathophysiology of bone abnormality may assist to further explain patients' symptoms and benefit in future management.

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REFERENCES

1. Bance M, Tysome JR, Smith ME. Patulous Eustachian tube (PET), a practical overview. World J Otorhinolaryngol Head Neck Surg 2019;11;5(3):137–42. doi: 10.1016/j.wjorl.2019.08.003.
2. Kim, Yi., Ryu, JS. Fibrous Dysplasia. Atlas of Nuclear Medicine in Musculoskeletal System. Springer, Singapore 2022:195-201. https://doi.org/10.1007/978-981-19-2677-8_16
3. Harisankar CN, Bhattacharya A, Bhadada SK, Kamaleshwaran KK, Mittal BR. An interesting case of polyostotic fibrous dysplasia: The "pirate sign" evaluated with Tc-99m methylene diphosphonate single-photon emission computed tomography/computerized tomography. Indian J Nucl Med. 2011;26(1):40-41. doi: 10.4103/0972-

3919.84613.
4. Couturier A, Aumaotre O, Gilain L, Jean B, Mom T, Andrй M. Craniofacial fibrous dysplasia: A 10-case series. Eur Ann Otorhinolaryngol Head Neck Dis 2017;134(4):229–35. doi: 10.1016/j.anorl.2017.02.004.
5. Suppiah S, Mohd Rohani MF, Zanial AZ, Ahmad Shahrir AD, Khairuman KA, Vinjamuri S. A Review on the Usage of Bone Single-Photon Emission Computed Tomography/Computed Tomography in Detecting Skeletal Metastases in the Post-COVID-19 Era: Is it Time to Ditch Planar and Single-Photon Emission Computed Tomography only Gamma Camera Systems? Ind J Nucl Med 2023;38(2):191-200. doi: 10.4103/ijnm.ijnm_142_22.