CASE REPORT

Pneumosinus dilatans multiplex associated with hormonal imbalance

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SUMMARY

Pneumosinus dilatans describes an abnormal dilation of one or more paranasal sinuses without radiological evidence of localised bone destruction, hyperostosis or mucous membrane thickening. Dilation of mastoid air cells also occurs rarely along with involvement of paranasal sinuses. This rare combination of unknown aetiology was reported in two cases in the literature and termed 'Pneumosinus Dilatans Multiplex' (PSDM). It is usually asymptomatic, and is detected incidentally on plain radiography, CT or MRI. If left untreated, it can further erode the bone leading to complications such as facial asymmetry, neurological disorders and pathological fractures. The aetiology of the condition remains obscure. Various hypotheses proposed are the presence of gas-forming microorganisms, spontaneous drainage of a mucocoele, the presence of a one-way valve, dysregulation of hormonal levels leading to a disturbance of osteoblastic and osteoclastic activity. This paper describes a case of PSDM possibly secondary to hormonal disturbance.

BACKGROUND

As to the best of our knowledge, the two reports of pneumosinus dilatans multiplex (PSDM) reported in the literature had mild facial asymmetry and mental retardation and the authors considered it to be a

Figure 2 Midline diastema.

syndromic condition.^{1 2} However, some authors have suggested that hormonal dysregulation could be a possible aetiological factor.³ However, there has been no case of pneumosinus dilatans (PSD) associated with hormonal alteration reported thus far in the literature.⁴

These findings of abnormally dilated sinuses should not be ignored as it can lead to complications such as intracranial meningioma,⁵ increased intracranial pressure, pathological fracture, exophthalmos, facial asymmetry and loss of vision.

CASE PRESENTATION

A 15-year-old boy reported for orthodontic consultation for closure of midline diastema (figures 1 and 2).

The patient was referred to the maxillofacial outpatient department as there was a suspicious lesion on the mandible as seen in the pretreatment orthopantomograph. He was totally asymptomatic. Intraoral examination revealed multiple missing

Figure 1 Facial symmetry.

Figure 3 Hyperpneumatization of the maxillary sinus extending to the anterior nasal spine along with a radiolucent lesion on the left mandible.

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Figure 4 (A) Osteolysis of the maxillary, temporal and mastoid region. (B) Abnormal dilation of the frontal sinus. (C) Abnormal dilation of the maxillary and ethmoid sinuses. (D) Hyperpneumatization of mastoid air cells. (E) Abnormal dilation of maxillary, sphenoid and ethmoid sinuses, as well as mastoid air cells.

premolars and retained primary teeth. There was no swelling and no alterations in consistency and texture on the left mandible.

INVESTIGATIONS

A comprehensive evaluation of the orthopantomography, revealed a 6×3 cm radiolucency on the left body of the mandible and

bilateral hyperpneumatisation of the maxillary sinus extending to the nasal spine which led to do further investigations (figure 3).

A multidetector CT revealed an overall osteolysis and abnormal pneumatization of all paranasal sinuses and air cells (figure 4A–E).

Blood investigations revealed an increase in parathyroid hormone levels (PTH 125 pg/mL) and alkaline phosphatase levels 275.2 IU/L associated with vitamin D deficiency (25-OH vitamin D 15.65 ng/mL), however, serum calcium and phosphorous levels were within normal limits.

A series of radiographs were taken to rule out involvement of axial skeleton and the findings were unremarkable. The patient was referred to the department of endocrinology for the management of hormonal imbalance.

Biopsy was performed for the lesion on the left body mandible. A bony window was created on the buccal cortex but no lining epithelium/soft tissue was found except for a hollow cavity which was quite similar to a solitary bone cyst.

DIFFERENTIAL DIAGNOSIS

All expansile lesion of sinonasal tract.

TREATMENT

During the biopsy procedure bleeding was induced followed by wound closure as this is the recommended treatment for solitary bone cysts (figure 5).

Vitamin D supplementation was prescribed as he was diagnosed with vitamin D insufficiency.

OUTCOME AND FOLLOW-UP

Periodic follow-up was conducted with relevant investigations. The patient showed signs of normalising PTH levels and vitamin D at a 3-month review.

DISCUSSION

Hwang *et al*¹ defined PSD as a localised abnormal dilation of one or more paranasal sinuses without radiological evidence of localised bone destruction, hyperostosis or mucous membrane thickening.¹

In the past, various terms have been used for this pathology, such as pneumocoele, pneumatocoele, pneumosinus frontalis,

sinus hypertrophy, aerocoele, hyperpneumatisation, sinus ectasia and sinus blistering. It was Benjamins⁶ in 1918 who distinguished between PSD and pneumocoele based on bony wall integrity. PSD and pneumocoeles can arise in any of the paranasal sinuses, and can be bilateral.

Teh *et al*,⁷ in a literature review, mentioned that the mean age of presentation was 27 years (range, 9–62 years), with a slight male predilection. The authors identified a total of 36 reported cases involving the maxillary sinus of which 2 cases also involve the mastoid air cells.^{8–9} The abnormal dilation of all paranasal sinuses, along with mastoid air cells, has been termed as PSDM.⁸ We present the third PSDM case in the literature that involves all paranasal sinuses and bilateral mastoid air cells.

The aetiology of PSD remains unknown. They have been described as primary or secondary. The proposed aetiologies of primary cases include a one-way valve effect,⁸ gas-forming organisms,¹⁰ spontaneously discharged mucocoele,¹¹ trauma¹² and dysregulation of hormones. However, there is no report of any hormonal alteration so far. In our case, the patient had vitamin D insufficiency which led to a decrease in serum calcium and phosphorus levels. This in turn would have led to the increase in parathyroid hormone levels which would have resulted in absorption of calcium from bones to normalise the serum calcium levels. PSD has been found to be associated with Melnick-Needles syndrome, Klippel-Trenaunay-Weber syndrome, Sturge-Weber syndrome, meningioma, arachnoid cysts, cerebral hemiatrophy, fibro-osseous disease and port-wine stain.¹³

PSD may present as a slowly progressing deformity that is asymptomatic. But if symptomatic, its clinical presentations include different types of facial deformity, impairment in visual acuity, headache and nasal obstruction. The typical presentation of PSD of frontal sinus includes frontal bossing and prominence of supraorbital ridge.¹⁴

Diagnosis is made by clinical examination along with confirmation by radiography (plain radiographs or CT). If diagnosis is made on plain radiography, additional cross-sectional imaging such as MRI or CT may be needed.

The treatment for PSD is based on the symptoms and facial deformity. Symptomatic patients need surgical intervention. Surgical options for maxillary PSD includes creation of a nasoantral window either by Caldwell-Luc operation¹³ or endoscopically,¹⁵ antral wall turnover (circumferential osteotomy of the convex maxillary sinus followed by removal of sinus membrane and then reinsertion of the bony fragment inside out), greenstick downward fracture and electrical burring.⁹ In cases of PSD of the frontal sinus the surgical treatment involves osteotomies followed by recontouring.¹⁴

Figure 5 Radiolucent lesion on the left mandible.

Learning points

- ▶ Abnormal dilation of all paranasal sinuses along with mastoid air cells has been termed as pneumosinus dilatans multiplex (PSDM).
- ▶ The proposed aetiologies include a one-way valve effect, gas-forming organisms, spontaneously discharged mucocoele, trauma and dysregulation of hormones.
- ▶ PSDM can lead to facial deformity, impairment in visual acuity, headache, nasal obstruction and frontal bossing.
- ▶ The treatment for pneumosinus dilatans is based on the symptoms and facial deformity. Symptomatic patients need surgical intervention.

Competing interests None.

Patient consent Obtained.

Provenance and peer review Not commissioned; externally peer reviewed.

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