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CASE REPORT

Radiographic manifestations of the temporomandibular joint in a case of Proteus syndrome

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Proteus syndrome is a rare disorder with progressive asymmetrical and disproportionate overgrowth of various tissues of the body. The syndrome is characterized by a wide range of malformations, including craniofacial deformities. Extraoral examination revealed several of the classical craniofacial features of Proteus syndrome: pronounced hemifacial hypertrophy, macrodactyly and hyperostosis. Intraoral examination revealed a high arched palate and gingival hyperplasia. Other findings were unilateral enlargement of the tongue, alveolar growth and dilaceration of the roots of the teeth. There were severe degenerative changes and deformities in the left temporomandibular joint but the oversized condyle was asymptomatic; there was no pain, limitation and deviation at mouth opening. Treatment was not necessary owing to the asymptomatic situation but periodic follow-up with clinical and radiographic examination was considered. The aim of this article is to describe the radiographic manifestations of an asymptomatic condyle malformation and other craniofacial, oral and dental findings in a 33-year-old female patient with known Proteus syndrome.
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Keywords: Proteus syndrome; temporomandibular joint; panoramic radiography; computed tomography

Introduction

Proteus syndrome (PS) is a rare disorder of patchy or mosaic post-natal overgrowth of unknown aetiology.[1] This syndrome was first delineated by Cohen and Hayden[2] in 1979, who reported a newly recognized disorder in two patients. 4 years later, Wiedemann et al[3] also reported four children with similar findings and named this condition after the Greek sea god Proteus, who could change his shape at will to avoid capture.

PS is characterized by asymmetrical and disproportionate overgrowth of connective tissue, the skeleton, blood vessels and central nervous system, but it can affect any tissue.[1] The syndrome affects more males than females.[4,5] One of the most common complications in patients with PS is deep venous thrombosis and pulmonary embolism.[1,5,6]

Diagnosis can be difficult because PS can affect various tissues of the body. The manifestations of the syndrome are highly variable and many are also found in other overgrowth syndromes.[7,8] The disorders most commonly confused with PS are Klippel–Trénaunay syndrome and hemihyperplasia–multiple lipomatosis syndrome.[9] The diagnosis depends heavily on clinical evaluation and imaging.[7] Diagnostic criteria for PS have been published by Biesecker et al[9] and Turner et al.[5]

Although several cases about the general clinical and genetic findings of PS have been reported in the literature, few reports have been published describing the dentomaxillofacial manifestations,[8,10–17] and the radiographic findings of the temporomandibular joint (TMJ) in PS have not been described in detail. Our case report presents the radiographic manifestations of an asymptomatic condyle malformation and also other

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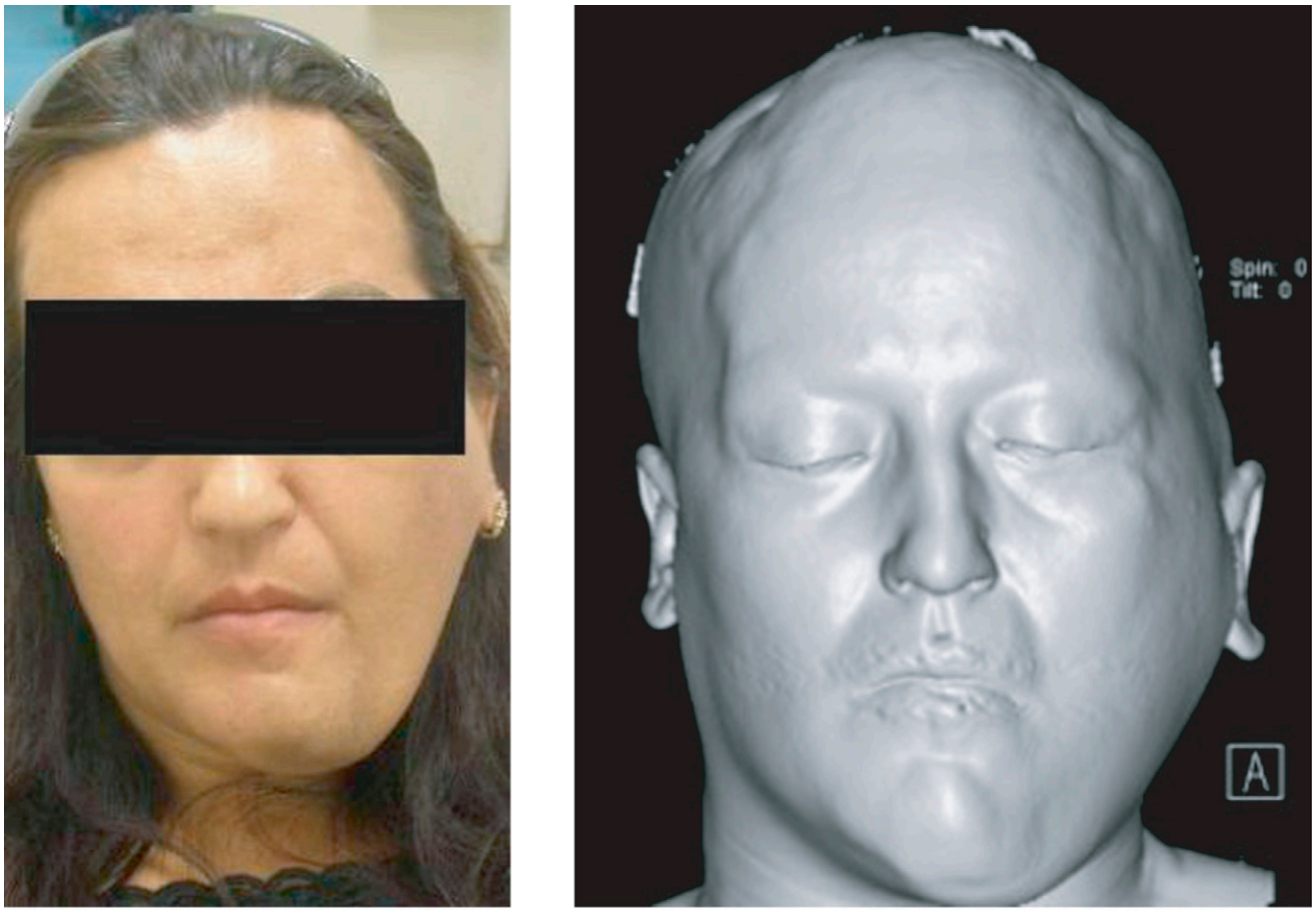

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Figure 1 (a) Extraoral photograph of the patient showing the marked facial asymmetry. (b) Volume rendered shaded surface display reformat of the three-dimensional head CT reveals hypertrophy and enlargement on the left side of the face

craniofacial, oral and dental findings in a female patient with known PS.

Case report

A 33-year-old female patient with known PS was referred to our department for dental examination. Informed consent was obtained from the patient. There was no family history of similar disorders. On examination, she had pronounced hemifacial hypertrophy of the left side (Figure 1a,b). Macrodactyly was present on the left first and middle fingers (Figure 2). The significantly enlarged index finger of the left hand was amputated 10 years before and multiple exostoses in the parietal, occipital, temporal and frontal bone were also surgically excised. She had abundant hair growth in her chin and cheek area on the affected side. There were no skin lesions on the face. Limitation and marked deviation were not observed at mouth opening. Intraoral examination showed the high arched palate and enlargement of the left mandibular posterior alveoler bone. Most of the teeth on the affected side were extracted owing to periodontal disease. Gingival hyperplasias were present in the patient, especially on the affected side. There was unilateral enlargement of the tongue on the same side (Figures 3 and 4).

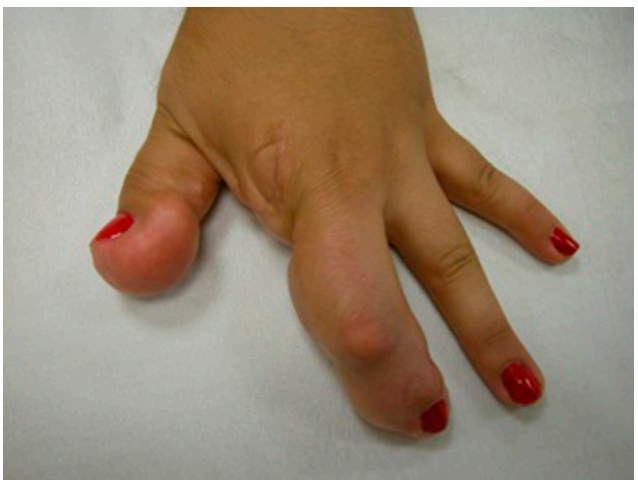

Figure 2 Photograph of the patient's left hand showing macrodactyly of the first and middle fingers. (Index finger has been amputated because of macrodactly)

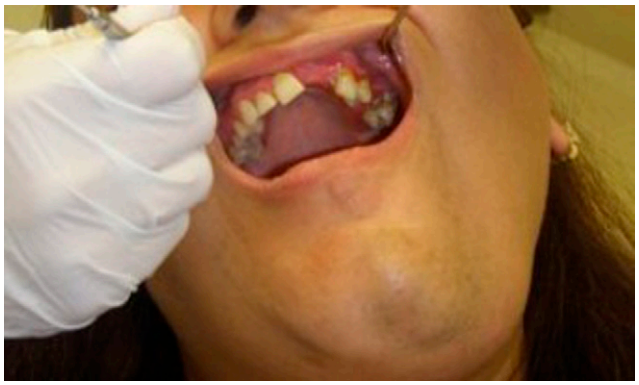

Figure 3 Intraoral view showing gingival hyperplasia and the high arched palate

A panoramic radiograph revealed mandibular hemihypertrophy of the left side. There was an excessive generalized growth of the left mandibular body and ramus. Hyperostoses in the left condyle and mandible were observed. There was coarse trabeculation of the left angulus mandible. The tuber maxilla of the affected side was also enlarged. More alveolar bone loss in the maxillary left side than the right side was detected. Dilaceration of the roots of both maxillary left canine and mandibular right central incisor was recorded. The panoramic radiograph also showed the left condyle malformation and wide sclerosis in the glenoid fossa (Figure 5). In addition to the panoramic radiograph, the open–closed lateral radiograph of the TMJ, which was obtained using the TMJ-specific program of the panoramic device, was also taken. This radiograph showed a significant limitation of movement of the condyle on the affected side, whereas there was anterior excursion of the unaffected condyle beyond the articular eminence (Figure 6). In order to obtain a more detailed image of the left TMJ morphology, CT was performed obtaining the sagittal and axial slices. CT images also demonstrated extensive degenerative changes of the left TMJ with pseudoarticulation of the coronoid process anteriorly. This pseudoarticulation was due to the hypertrophic changes of the temporal bone and mandibular condyle (Figure 7a,b). Additionally, CT images of the head revealed new bone formation in the occipital and temporal region.

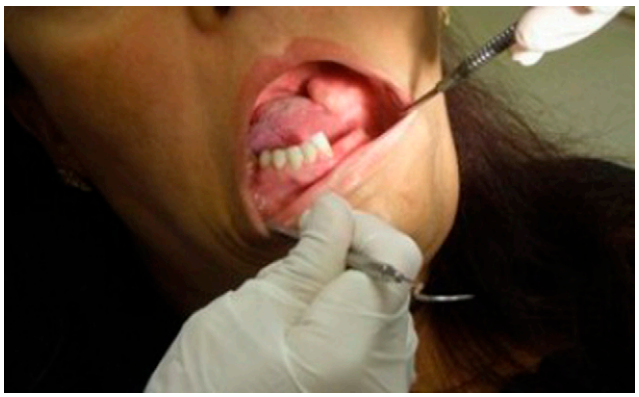

Figure 4 Intraoral view showing unilateral enlargement of the tongue and alveolar bone overgrowth on the posterior part of the left mandibular

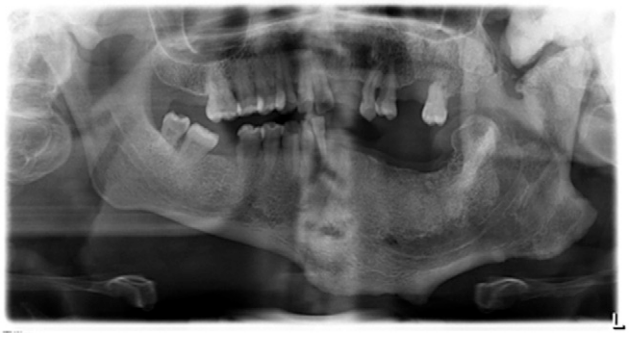

Figure 5 Panoramic radiograph showing mandibular hemihypertrophy of the left side and the left condyle malformation and wide sclerosis in the glenoid fossa

Discussion

PS is a rare complex disorder characterized by asymmetrical and disproportionate overgrowth of various tissues of the body.[4] Since PS is progressive in nature, the craniofacial distortion becomes more evident with age.[18] The disease may slow or stabilize during early adolescence.[7,11] The disease process is not usually apparent at birth; the onset of overgrowth commonly occurs in childhood.[7] In our female patient the onset of asymmetrical overgrowth started at the age of 7 years.

The clinical manifestations are highly variable, because of multifocal overgrowth affecting any tissue of the body.[1,3–5,7,9,18,19] In this case, extraoral examination revealed several of the previously reported craniofacial features of PS: pronounced hemifacial hypertrophy, macrodactyly and hyperostosis. Both the left tuber maxilla and the mandible were affected. Craniofacial hyperostoses are common in PS.[19] They may occur in the cranium, nasal bones, external auditory meatus, alveolar ridge, maxillary and mandibular bone.[1,5,9,14,17–19] In the presented case, hyperostoses in the alveolar bone, condyle and mandible were observed on the affected side. Multiple skull hyperostoses involving the parietal, occipital, temporal and frontal bone were removed surgically 10 years before. The tissue overgrowth may stabilize after adolescence,[7,11] but in our patient new bone overgrowth has been radiographically observed in the occipital and temporal region following the surgery.

Dental and intraoral manifestations of PS have been described in the literature.[8,10–13,15–17,19] A high arched palate and unilateral enlargement of the tongue on the affected side were found in our patient. Most of the teeth on the left side were extracted owing to periodontal disease. There were only three teeth on the affected side,

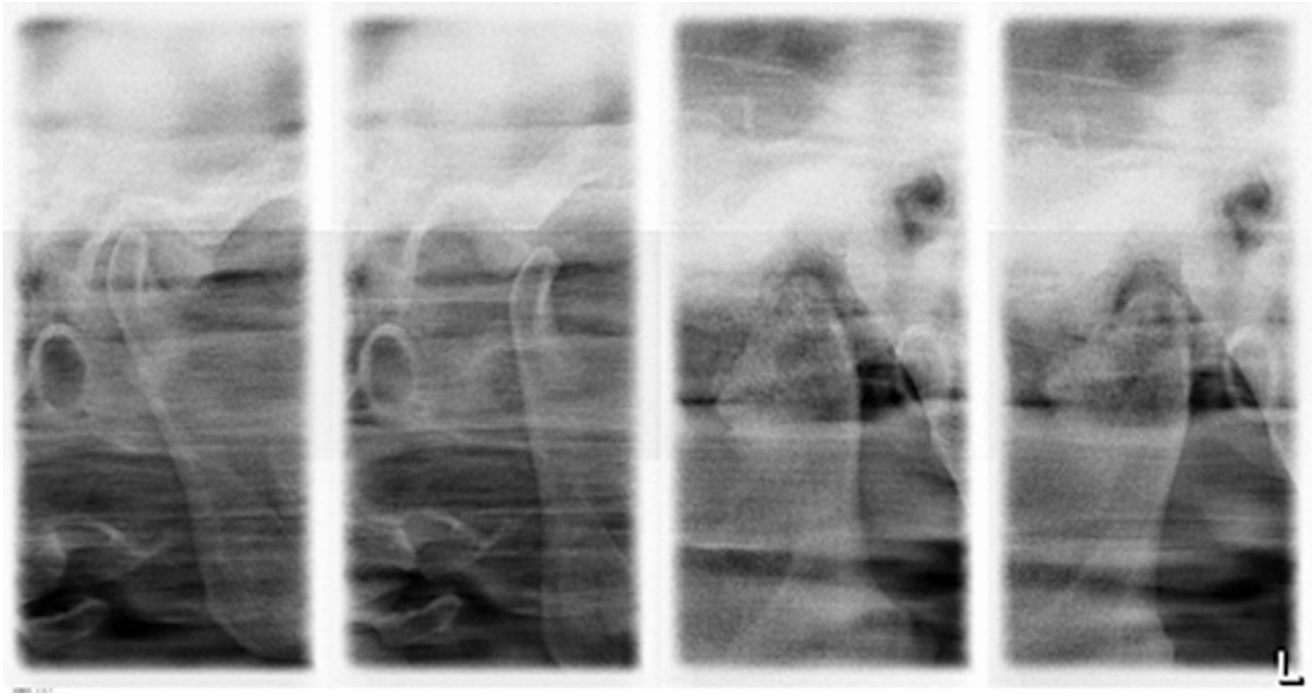

Figure 6 The open–closed lateral radiograph of the temporomandibular joint showing a significant limitation of movement of the condyle on the affected side

and both gingival hyperplasia and alveolar bone destruction were detected. There was only dilaceration of the roots of both maxillary left canine and mandibular right central incisor; other dental anomalies and intraoral findings were not observed.

Skeletal developmental malformations are the most frequent finding in PS. The disproportionate overgrowth seen in PS is commonly associated with irregular and disorganized bone, including hyperproliferation of osteoid with variable calcification, resulting in abnormal bony edges, abnormally calcified connective tissue and

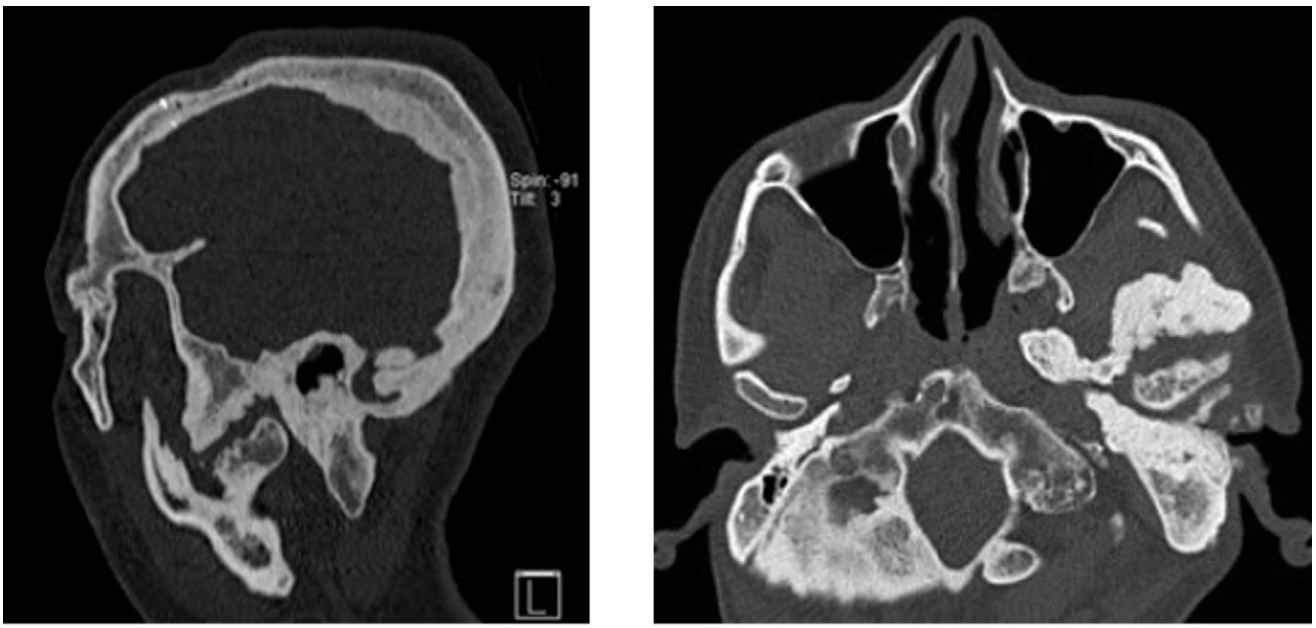

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Figure 7 (a) Sagittal multiplanar reformat demonstrating extensive degenerative changes of the left temporomandibular joint with pseudoarticulation of the coronoid process anteriorly. (b) Axial image of the three dimensional CT reveals hypertrophic changes of the temporal bone and mandibular condyle with accompanying degenerative changes

bony invasion of joint spaces, eventually resulting in immobility of the affected joint.[5,7] Unilateral condylar hyperplasia has been mentioned in the literature,[8,12,15] but the unilateral condylar hyperplasia characteristics have not been described. In the present case, the panoramic radiograph showed the left condyle malformation and wide sclerosis in the glenoid fossa. In addition to the panoramic radiograph, the open–closed lateral radiograph of the TMJ and CT images were obtained for a more detailed image of the condylar region. Severe degenerative changes, jagged bone edges and deformities in the left TMJ were the most striking radiographic features. Wide sclerosis owing to probable abnormally calcified tissue in the left glenoid fossa was also observed. The open–closed lateral radiograph of the TMJ also provided additional information about the movement of the TMJ. The radiographic examination revealed minimal movement of the left condyle whereas there was anterior excursion of the right condyle beyond the articular eminence.

A radiological examination is an essential part of the diagnosis and management of TMJ disease. Panoramic radiography is a useful screening technique for condylar abnormalities, but accurate evaluation of the TMJ has been difficult owing to the superimposition of other structures in conventional radiographs.[20] In addition the panoramic view does not provide adequate examination of the shape of the condyle because of the distorted view of the joint.[21] CT provides precise imaging of TMJ anatomy without superimposition and distortion. CT is the ideal imaging choice for evaluating hard tissues.[20]

There are no completely satisfactory orthopaedic procedures to treat these lesions—the therapy should be symptomatic and a multidisciplinary approach should be taken owing to the syndrome's complexity.[15,22] The aim of the treatment is minimization of the disability, and contributions can be made by plastic, dental and orthopaedic surgeons and physiotherapists.[16] In our case, although severe degenerative changes and deformities in the left TMJ were radiographically observed, the oversized condyle was asymptomatic and there was no pain, limitation or deviation at mouth opening. For this reason, no treatment was applied to the patient and the clinical and radiographic follow-up was considered because of its progressive nature.

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