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內文：

Introduction

1. Progressive systemic sclerosis (PSS), also known as scleroderma, is a connective tissue disorder characterized by fibrosis of the skin and visceral organs.
2. The cause of PSS is immunologically mediated pathogenesis.
3. PSS occurs predominately in the adult but it can also be seen in childhood, females: males=3~4:1
4. The most common manifestations is microstomia that causes a limitation of opening the mouth.
5. In addition, salivary gland fibrosis causes xerostomia; these patients are more prone to dental decay.
6. In some patients, as loss of attachment gingival occurs, multiple area of gingival recession may be present.
7. Oral hygiene instruction (OHI) and preventive dentistry are recommended for these patients.
8. Occasionally slight bilateral erosion of mandibular condyle causes anterior open bite.
9. Bone resorption is caused by the increased pressure associated with the deposition of collagen.

Case Report

1. A 36-year-old female suffering from PSS with 9-year history of disease was referred to oral and maxillofacial surgery for extraction of her decayed posterior teeth.
2. MIO = 12 mm, mask-like face, perioral deep rhytides, pinched nose, facial telangiectasia, and atrophic ulcerated finger tips (sclerodactyly). Pain accompanying bone resorption is present in her fingers (painful digital ischemia).

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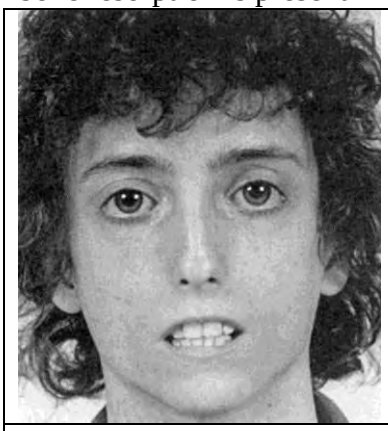
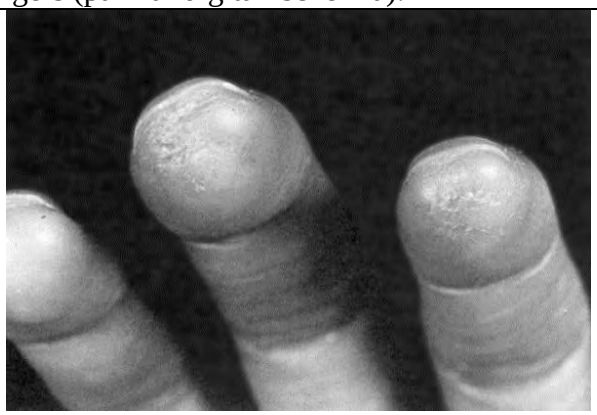
<tr><td>  </td><td>  </td></tr>
<tr><td>Small mouth with puckering of the lips, pinched nose</td><td>Tiny digital pitting scars resulting from digital ischemia</td></tr>
<tr><td colspan="2">(http://www.uptomed.ir/Digimed.ir/nelson-textbook-of-pediatrics-18th-edition/Nelson_Textbook_of_Pediatrics__18th_Edition/HTML/473.htm)</td></tr>
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3. Medical history: pulmonary fibrosis. Limitation in mouth opening, malformed

fingers, dry mouth, tight tongue, and reduced movement of the cheeks created problems in oral hygiene maintenance

4. Dental treatment such as posterior teeth restorations, fixed or removable prosthesis, was difficult to do in this patient because of restricted mouth opening.

5. Dental panoramic view relieved extensive bone resorption of bilateral mandibular angles and total left condylolysis.

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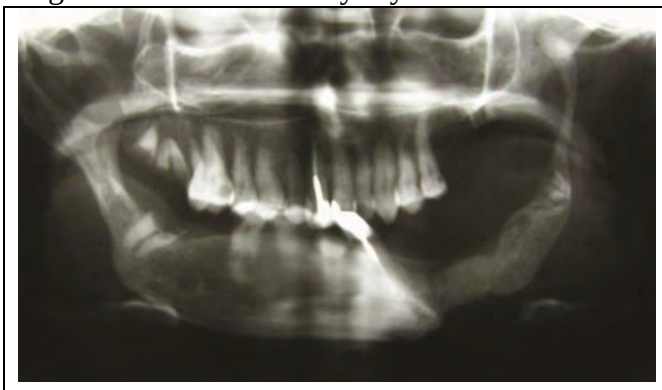
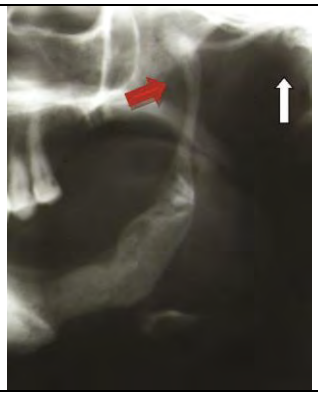
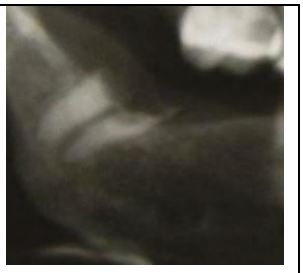
|  |  |  |
| Extensive bone resorption of bilateral mandibular angles and total left condylolysis | -Empty glenoid fossa (white )<br>-thin elongated coronoid process (red)<br>-left angle bone resorption | Resorption of the right mandibular angle and decayed remnant root tooth. |

6. There was a 1.5 cm vertical length difference between right and left coronoid processes. Patient has no history of jaw trauma or surgery.

7. Under local anesthesia, decayed roots were removed. Limitation in mouth opening and severe bone resorption made dental procedures difficult. Great caution is required to avoid iatrogenic jaw fracture.

Discussion

1. Scleroderma or PSS is an autoimmune disease characterized by gradual deposition of collagen in reticular dermis, gastrointestinal tract, heart, lung, and other organs

2. PSS most commonly occur during the third to fifth decades of life. PSS has a low prevalence (130 per million)

3. Bone resorption is an uncommon occurrence but recognized complication of long standing PSS

4. In maxillofacial regions resorption occurs in attachment area of masticatory muscles including masseter, temporal, lateral pterygoid, and anterior belly of digastrics muscle.

5. Mandibular bone resorption usually happens in mandibular angle area and is more likely to happen bilaterally.

6. Pain, preauricular tenderness, occlusal disturbance, TMJ sounds, and reduction of ramus height are common clinical features, but in our case despite huge bone resorption, there were no pain and tenderness.

7. Dental radiographic manifestations of the scleroderma patient include PDL widening, root resorption, and decayed posterior teeth similar to our case.

8. Patients with PSS exhibit an increased prevalence of external root resorption. Root canal therapies of the teeth with external root resorption require special consideration.

9. In this case we should consider concurrent secondary sjogren syndrome. In addition, there is an increased prevalence of oral candidacies in patients with

xerostomia because of reduction in saliva production.
10. In the present case, dry mouth was an important clinical feature; thus preventive dentistry is too important in scleroderma patients.
11. In this case, perimandibular connective tissue (PMCT) adaptation and presence of mandibular canines help in stabilization of three dimensional spatial positions of jaws.
12. Deposition of abnormal collagen in the tongue causes a firm and hypomobile tongue. The oral chief compliant in scleroderma patients is xerostomia, which causes difficulty in speaking.

| 題號 | 題目 |
|---|---|
| 1 | 關於謝格連氏症候群(Sjogren syndrome)，下列敘述何者正確 ？<br>(A) 它是一種第 IV 型的過敏反應<br>(B) 鏡下常見到淋巴上皮病灶(lymphoepithelial leision)，都應視為低惡性度早期的黏膜相關性的淋巴組織淋巴瘤(MALT lymphoma)<br>(C) 好犯中年男性<br>(D) 鏡下可以見到特別的病理變化稱為上皮肌上皮性島(epimyoepithelial island) |
| 答案(D) | 出處：Oral and Maxillofacial Pathology, 2009, 3th edition. p466-p470 |
| 題號 | 題目 |
| 2 | 下列何種情況不會造成口乾症(xerostomia)<br>(A) 謝格連氏症候群 (Sjogren syndrome)<br>(B) 糖尿病 (diabetes mellitus)<br>(C) 腹式呼吸法 (abdominal breathing)<br>(D) 服用 methyldopa 藥物 |
| 答案(C ) | 出處：Oral and Maxillofacial Pathology, 2009, 3th edition. p398 |