ΒΡΑΧΕΙΑ ΔΗΜΟΣΙΕΥΣΗ
SHORT COMMUNICATION

Ascher syndrome and chronic orofacial pain disorder

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Σύνδρομο Ascher σε ασθενή με χρόνιο στοματικό και προσωπικό άλγος

Περίληψη στο τέλος του άρθρου

Key words: Ascher syndrome, Chronic orofacial pain, Fibrous dysplasia

Ascher syndrome is a rare condition, characterized by blepharochalasis and double lip with or without non-toxic goitre, first described by Ascher in 1920.[1] Blepharochalasis is an unusual disorder classified by Fuchs in 1896.[2]

It is clinically manifest by recurrent episodes of bilateral upper eyelid edema, generally non-pitting, free of pain, non-erythematous and with increased upper eyelid skin laxity, prolapsed or herniated fat and periorbital skin discolouration or hyperpigmentation.[3–7] Historicaly, this condition was previously described in 1909 as a trophoneurosis.[8]

The double lip consists of two segments and often the inner part, or pars villosa, sags beneath the outer part of the lip, the pars glabrosa.[1,9] The pars villosa may be veiled by the pars glabrosa when the patient relaxes, making the diagnosis more difficult. In such cases, smiling usually reveals the sign of the double lip.

Thyroid gland involvement is not a constant component of the syndrome.[5,7,10–13] In approximately 10% of cases, non-toxic goitre is present.[9,10,13]

Chronic orofacial pain is a common condition that can be subdivided into chronic temporomandibular joint disorder (facial arthromyalgia), atypical or chronic facial pain, atypical or chronic odontalgia and oral dysesthesia.[14]

Pain is an unpleasant sensory and emotional experience associated with actual or potential tissue damage.[15] This typifies the medico-dental entities of the chronic orofacial pain disorders. Chronic temporomandibular joint disorders are common conditions affecting 33% of the population,[16] characterized as unilateral or bilateral pain in the temporomandibular joint and its associated craniofacial musculature, with additional ear symptoms such as a sensation of fullness and popping.[14,17] The most common symptoms are pain on palpation of the joint and muscles of mastication with or without reduction in the extent of mouth opening and mandibular movements, clicking or grating sounds in the joint.[18]

Fibrous dysplasia, a benign fibro-osseous condition, has not been previously associated with either chronic facial pain or Ascher syndrome.

CASE REPORT

A 57 year-old lady was referred to the Maxillofacial and Oral Surgery Department suffering from recurrent swellings of the mid-face and eyes, chronic bilateral temporomandibular joint dysfunction (TMJD), left maxillary tenderness, fatigue and depression. The medical history revealed diabetes mellitus, recurrent sinusitis, hay fever and a mild stroke.

On clinical examination, sagging of the upper eyelids (fig. 1) and double folding of the upper lip (fig. 2) were observed. The temporomandibular joints were tender on palpation and there was bilateral clicking of the joints and deviation of the mandible to the left on mouth opening. Intra-oral examination revealed marked enlargement of the maxillary tuberosity.

Fibrous dysplasia was diagnosed using plain radiographs (fig. 3) and CT examination followed by incisional biopsy for histopathological confirmation. The fibro-osseous lesion consisted of bone expansion with preservation of the cortex but a typical ground glass appearance consistent with the diagnosis. The condition involved the left maxillary alveolus and bilateral temporal bones at the junction with the lateral border of the orbits. The patient was reassured of the benign nature of the condition.

The laboratory examination revealed no thyroid gland involvement (TSH: 4.6 mU/L, free T_4: 12.5 pmol/L and free T_3: 5.4 pmol/L). The renal blood chemistry was in the normal

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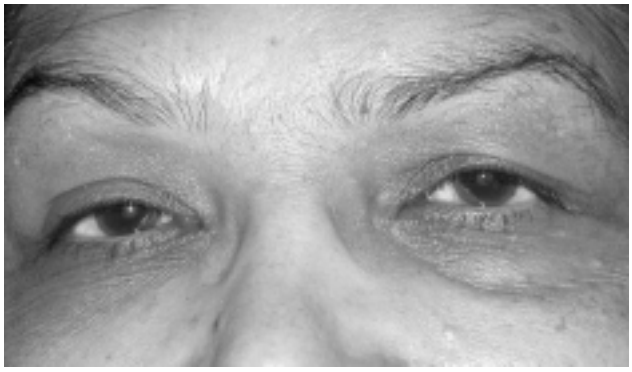

Figure 1. Blepharochalasis is observed, with upper eyelid sagging and periorbital hyperpigmentation. The condition is more prominent on the left side.

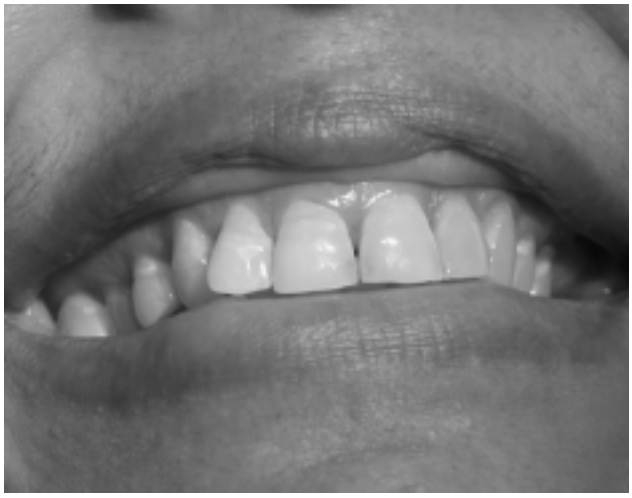

Figure 2. The upper double lip is observed only when the patient smiles.

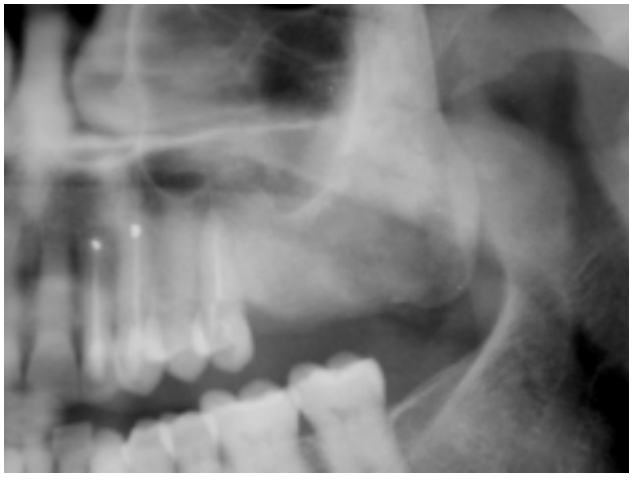

Figure 3. Dental panoramic tomography showing fibrous dysplasia of the left maxillary alveolus.

range (Na: 140 mmol/L, K: 4.5 mmol/L, urea: 3.2 mmol/L) with slightly decreased creatinine clearance: 53 umol/L. In the liver and bone evaluation the alkaline phosphatase was slightly increased at 132 IU/L. The full blood count was normal. The diagnosis of an incomplete form of Ascher syndrome was established by the history, the clinical examination and laboratory investigations.

COMMENT

The various suggestions regarding the etiology of Ascher syndrome remain inconclusive. Several cases have been reported in the non-English literature, making the study of the complete data relatively difficult. Since the first description of Ascher syndrome only about 50 cases have been reported.[10] Literature review shows no significant sex-prevalence for the syndrome and the most common age presentation ranges from 11 to 26 years.[1,5,7,9–11,13,19] The usual onset of appearance of Ascher syndrome is between 10 and 18 years of age and it persists throughout life.[5] Features are usually first observed before the age of 20 in 80% of cases.[13] The cases of two men, 56 and 75 years old, have also been reported, illustrating that if the facial esthetics and function are not significantly affected the symptoms may remain undiagnosed for a long period of time.[4,11] In this particular case study, the lady was undiagnosed until the age of 57 years.

Blepharochalasis is refractory to antihistamines and corticosteroids, which reveals its lack of relation to allergic, immunological mechanisms.[10] Histopathologically, the epidermal thickness of the eyelid skin varies from normal to moderately atrophic, with numerous capillaries observed.[3,4] This differentiates the condition from dermatochalasis in the elderly caused by dermal atrophy and folding of the upper eyelid. The ophthalmologic assessment of blepharochalasis in severe cases may show ectropion or entropion of eyelids and erosion of the upper portion of sclera by the resultant trichiasis.[7,11]

In addition to upper eyelid blepharochalasis, the double folding of the upper lip includes one of the diagnostic standards for the incomplete form of Ascher syndrome. The name procheilia has been used to express this condition,[1] but the more commonly observed double lip seems more appropriate terminology. The prominent histological findings of upper double lip are minor salivary glands and a mixed inflammatory cell infiltration.[11]

The upper double lip can be difficult to distinguish when the upper labial mucosa is only evident when the patient smiles.[6,19]

The enlargement of the thyroid gland is non-toxic in the complete form of this syndrome and can be evaluated by scanning, after administration of ^{131}I, or using thyroid gammagraphy.[11,19]

The possible relationship of endocrinology and multiple system involvement with blepharochalasis and double lip has been suggested.[20–22] The normal values of 17-ketosteroid and estrogen with normal adrenal function reported in other studies do not confirm these findings.[7,13,19] An incomplete case of Ascher syndrome with bilateral cryptorchidism has been discussed but this association seems coincidental.[13] The relation of the rheumatic diseases and Chotzen-Saethre syndrome with Ascher syndrome has not been verified.[19,23]

Blepharochalasis and double lip is a benign condition, which requires surgical intervention only with severe ophthalmic involvement or when the patient is esthetically compromised.[3,6,10]

In conclusion, Ascher syndrome is an infrequent clinical anomaly of unknown origin. The presentation of chronic orofacial pain and craniofacial fibro-osseous lesions in a patient with Ascher syndrome seems to be coincidental in the absence of supportive information. Although it did not affect the management of this case, it did explain the intermittent peri-orbital swelling, which served to reassure the patient and to eliminate the symptom as a separate entity from the fibrous dysplasia and chronic temporomandibular joint disorder.

ΠΕΡΙΛΗΨΗ

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Το σύνδρομο του Ascher είναι μια σπάνια κλινική οντότητα, που χαρακτηρίζεται από (α) άνω διπλό χείλος, (β) βλεφαροχάλαση άνω βλεφάρων και (γ) συνύπαρξη μη τοξικής βρογχοκήλης, στο 10% των περιπτώσεων. Στη συγκεκριμένη εργασία χρησιμοποιήθηκε κυρίως η Αγγλική βιβλιογραφία μέσω του ηλεκτρονικού συστήματος Medline. Η παθογένεια του προαναφερθέντος συνδρόμου δεν έχει διευκρινιστεί. Ο πιθανός συσχετισμός με νοσήματα του ενδοκρινικού συστήματος ή πολλαπλών συστημάτων έχει αναφερθεί στη βιβλιογραφία, αλλά προς το παρόν παραμένει άλυτος γρίφος. Η παρουσία του συνδρόμου Ascher σε συσχετισμό με χρόνιο στοματικό και γναθοπροσωπικό άλγος, καθώς και ινώδη δυσπλασία του κρανιακού συμπλέγματος, δεν έχει αναφερθεί σε προηγούμενες μελέτες. Η πιθανότητα της σχέσης αυτής αναλύεται στην παρούσα εργασία.

Λέξεις ευρετηρίου: Ινώδης δυσπλασία, Σύνδρομο Ascher, Χρόνιο γναθοπροσωπικό άλγος

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