FRI0203 PULMONARY INVOLVEMENT IN JUVENILE SCLERODERMA

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Background Scleroderma in children manifests more frequently in the form of localised disease than systemic one. This disorder is characterised by the presence of hard and tight skin (linear, localised and generalised morphea); systemic sclerosis, besides the skin changes it manifests the internal organ involvement.

Objectives The aim of this report was an assessment of the frequency and nature of lung lesions in children with scleroderma.

Methods The study comprised 32 non selected pts, aged from 4 to 18 yrs., 16 suffering from localised and 16 from systemic scleroderma. The disease duration ranged from 6 months to 14 yrs. The relation between clinical symptoms, x – ray findings and functional pulmonary tests were performed.

Results Clinical and/or radiological signs (cough, dyspnoe, interstitial thickening) were found in 9 pts. The results of the functional investigation showed an impairment of the pulmonary function in 80.6% of pts with systemic sclerosis and 50% – with localised scleroderma, (total 65.6%). A decrease in DLCO (in 1/3 of pts in association with restrictive ventilatory failure) belonged to the most frequent of the lung damages.

Conclusion

- Pulmonary involvement is frequent, however not a main feature in the course juvenile scleroderma.
- Abnormalities in functional tests were often the only manifestation of the respiratory system involvement.
- Lung functional disturbances may be independent of clinical features, muscular on skin involvement.
- Respiratory functional tests are helpful in estimation of systemic scleroderma in the cases manifesting only the clinical form of localised one.

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FRI0204 TRIGEMINAL SENSORY NEUROPATHY IN SYSTEMIC SCLEROSIS

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Background

Objectives Systemic sclerosis (SSc) is one of the few somatic disorders which induce isolated or combined trigeminal sensory neuropathy (TSN).

Methods A screening of 101 patients with SSc hospitalised at the Institute of Rheumatology over a period of six months in 2000 revealed that eight patients (7.9%) had TSN (all of them were females aged 42–69 with a clinical history of SSc of 1 to

12 years). In two cases, SSc was associated with the Sjögren's syndrome; one case was associated with polymyositis. All the patients with TSN underwent rheumatological and neurological examinations including the recording of blink reflex.

Results The main clinical manifestation of TSN was bilateral asymmetric numbness in all the three divisions of the trigeminal nerve, especially in the perioral area. Five patients had qualitative taste disorders including «oral astereognosis», i.e., monotypical sensations in the oral cavity on taking up foods of different kinds and tastes. In two cases, TSN was manifested as constant and constant or «neuralgia-like» facial pains. Irrespective of remissions (which could be spontaneous), in each case facial pain was one of the most significant manifestations of SSc. All the patients had disturbances in surface sensitivity (hyper- or hypoesthesia) in the distal parts of the upper and lower extremities. In three cases, early symptoms of TSN (e.g., facial paraesthesia, numbness and hyperesthesia) developed simultaneously with the changes in the skin colour (especially of fingers), skin oedema, arthralgias and malaise. In the rest of the patients, TSN developed within 6 months to 3 years after the Raynaud's syndrome, sclerodactyly and other manifestations of SSc. The blink reflex records revealed modest prolongation of ipsilateral R1 responses. Specific therapy of TSN included administration of vasoactive drugs (e.g., pentoxifylline, vazaprostan) and group B vitamins. The elimination of facial pain presented a problem. The use of NSAID resulted in short-term attenuation of pain; in one case, topical application of ibuprofen (in the form of a cream) was more effective. Patients with paroxysmal manifestations of pain received additionally carbamazepine (300–400 mg/d). The efficiency of physio- and reflexotherapy in patients with acute facial pain syndrome is open to question.

Conclusion TSN is a typical neurological sign of SSc (with frequency 7.9% of cases), which is manifested as facial numbness, qualitative taste disorders including «oral astereognosis», pain and paraesthesiae. Most often, TSN develops within 6 months to 3 years after the appearance of other characteristic features of SSc. A combination of bilateral sensory disturbances in the facial area and distal parts of the upper and lower extremities suggests that TSN can be regarded as a partial form of generalised sensory polyneuropathy.

FRI0205 CLUES TO SYSTEMIC SCLEROSIS SINE SCLERODERMA

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Background There is substantial difficulty in the diagnosis of progressive systemic sclerosis where visceral involvement precedes cutaneous expression of the disease.

Objectives To describe clues to the diagnosis of progressive systemic sclerosis sine scleroderma (ssSSc).

Methods Report of 3 cases and review of the MEDLINE database concerning ssSSc.

Results Twenty five patients from the literature and 3 from our unit fulfilled the inclusion criteria, 13 females and 15 males, mean age 51.2(SD17.2) years. Conditions motivating the search for ssSSc were pulmonary fibrosis (44%), congestive heart failure (19%), esophageal dysfunction (7%), malabsorption (11%), nephropathy (11%), fever of unknown origin (3.5%) and seizures (3.5%). An overt undifferentiated connective tissue disease was present in 7 pts. Multisystem involvement became apparent