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TIETZE'S SYNDROME

REPORT OF A CASE WITH JUVENILE ONSET AND REVIEW OF THE LITERATURE*

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In 1921, Tietze first described a syndrome that consists of pain, tenderness and swelling of the costal cartilages and usually involves the second left costosternal or sternoclavicular joints[1]. This syndrome occurs in all age groups, but especially in the 2nd and 3rd decades of life. In some countries, where coronary heart disease is common and where people are very conscious of thoracic pain, most adults with this syndrome present themselves in the emergency room, convinced that they are about to have a heart attack. Only a few cases of this syndrome in the pediatric age group are to be found in the literature, and a case like our present one, of a patient who has not yet reached puberty, has rarely been reported[2-5].

Case Report

The patient, a thirteen-year-old boy, was first seen in the outpatient clinic of the Uludağ University Faculty of Medicine on 27 December 1979. Until the previous week he had been in good health, but then he began to have a sharp, intermittent pain over the precordial region. The pain was exacerbated by coughing and deep inspiration. He had been admitted to another hospital for a week, but, since the pain increased progressively and the treatment was apparently ineffective, he was transferred to our department.

Physical examination revealed a well-developed, apparently well-nourished child. Body temperature, pulse-rate and respiration-rate were normal, as were other physical findings. Blood pressure was 110/70 mmHg, radiological findings of the thorax were negative; when pressure was applied to the lower left part of the thorax there was pain; there was no local swelling, erythema or heat. Electrocardiograms were within normal limits. Hemoglobin was 13 gr/dl, the leukocyte count and the peripheral blood smear were within normal limits. Liver function tests were also normal.

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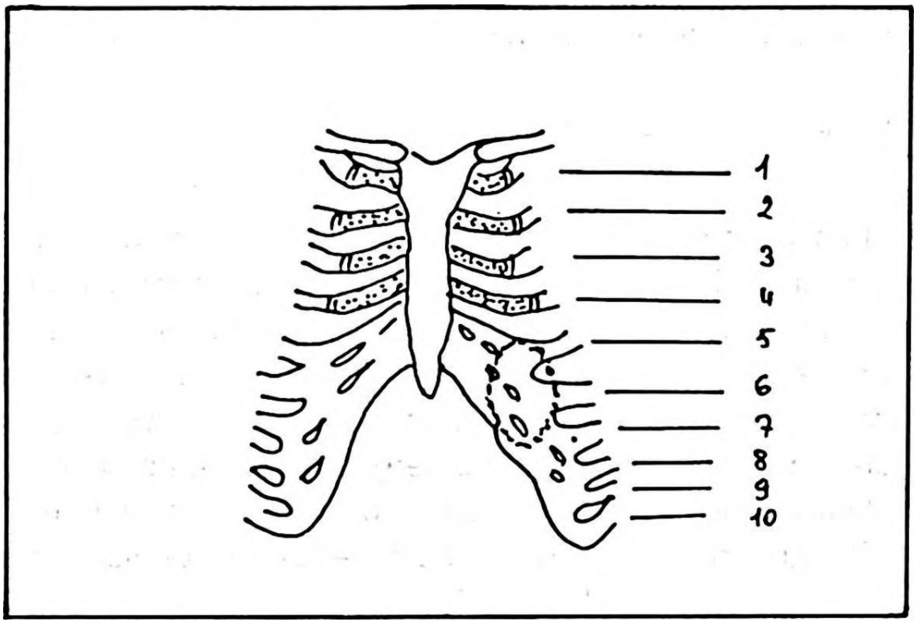
By the 8th day after admission, the pain had increased considerably; meanwhile a hard, fixed, tender, fusiform swelling had developed which measured approximately 3 cm by 5 cm and spread over the left 5th, 6th and 7th costal cartilages; there was no erythema in the area nor was there heat; the overlying skin moved freely and was neither edematous nor ulcerated (Figure 1). Radiological examinations were also normal at this stage.



On January 28, 1980, a surgical intervention was performed in the swollen region and a biopsy was taken. During this procedure there was no evidence of a tumor, inflammation or adhesion in the tissues overlying the cartilages. A small wedge of tissue was excised together with costal cartilage; this showed no evidence of tumor or inflammation. The operative incision healed normally.

Two weeks after the operation, the pain came back this time on the right side of the thorax. Analgesics were used, but they were effective only for a few hours. The patient was very irritable; a local injection of methyl prednisolone was effective for a reasonable length of time.

Discussion

The exact cause of Tietze's syndrome is unknown, but it is generally agreed to be mild trauma, particularly repeated trauma from coughing, respiratory diseases or sudden movements which produce small tears in the costal ligaments, that are responsible for this syndrome. It may not be a disease of cartilage at all, but of the surrounding ligaments. Many predisposing factors such as nutritional deficiencies, pulmonary tuberculosis, rheumatic or arthritic conditions have been suspected but these have not been proved to have an etiologic importance. External trauma has also been put forward as a precipitating factor, in some patients, by some clinicians[6-8].

The chief complaint in the vast majority of reported cases is pain, and this can radiate widely, thereby simulating intrathoracic or intraabdominal diseases. The pain, which is variable in intensity, may radiate as far as shoulders and arms, and it is often intensified by sneezing, coughing, inspiration, bending and exertion[9]. Examination discloses a nonfluctuant, firm, tender, bulbous swelling on the chest. A few patients have given histories of such a swelling being present for three to five years. A prolonged course is the rule[8]. Systemic manifestations are usually absent or minimal[3,9].

The roentgenographic studies are generally normal[2,6,8] but occasionally there is increased calcification on the affected side[6]. The chest X-ray is important since other diseases can thus be eliminated. Routine laboratory investigations have been reported as normal in several cases[2,8,9].

Positive diagnosis can be made only when a biopsy specimen shows an essentially normal costal cartilage[8,10]. A surgical biopsy is necessary to rule out many specific lesions, which may cause a swelling of the cartilages. These include benign and malignant neoplasms like chondroma, osteochondroma, multiple myeloma, osteogenic sarcoma, Ewing's tumor, Hodgkin's disease and various metastatic tumors, in particular carcinoma of the breast and the lung and also certain pyogenic infections, such as osteomyelitis of the rib and metastatic bacterial osteochondritis. Other lesions in costal cartilages such as those resulting from tuberculosis, actinomycosis and brucellosis might also show similar symptoms[8,11,12].

The question may arise as to whether this syndrome is not simply a painful chondroma, since the histological distinction between chondroma and normal cartilage is sometimes a difficult one. However several differences are apparent: the typical chondroma is lobulated, whereas no lobulation has been reported in Tietze's syndrome. A chondroma grows steadily and may reach large proportions, whereas the lesions of Tietze's syndrome never constitute more than a slight swelling which is capable of spontaneous regression[8].

Although the terms "Tietze's syndrome" and "costochondritis" are often used interchangeably, Tietze's syndrome is associated with local swelling whereas costochondritis is not. Consequently Tietze's syndrome is easier to recognize.

The treatment of this syndrome is symptomatic. In some cases local heat therapy and salicylates are helpful. Oral anti-inflammatory drugs are not usually effective. X-ray therapy has been tried but found to be of little or no benefit[13]. Local injection of procaine has also been tried but has brought little or no relief. Methyl prednisolone acetate at a dose of 40 mg per injection has had no adverse side effects and has been beneficial in most cases[7,13]. Partial or complete resection of the involved cartilage has been carried out in refractory cases and there has been no recurrence of the condition in such patients[11].

Summary

In this paper the case of a 13-year-old child with Tietze's syndrome is presented. Tietze's syndrome is characterized by pain in the anterior chest wall and by local swelling of the costal cartilages for no apparent reason. The clinical course is benign but may be prolonged over three to five years. This syndrome is very rarely seen in children.

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