REVIEW ARTICLE

TIETZE SYNDROME REVISITED

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ABSTRACT

Tietze syndrome is a rare condition. It is sometimes classified as a seronegative arthropathy. Its importance lies in the fact that it is often confused with costochondritis and ischaemic heart disease (much more common conditions) even though it is a distinct clinical entity. This article reviews the current concepts regarding this disease.

KEYWORDS: Tietze syndrome, chest wall tenderness, chest wall swelling

INTRODUCTION

Tietze syndrome is a rare condition but its importance lies in the fact that it comes in the differential diagnosis of chest pain and also because it is often confused with costochondritis which is a closely related but much more common condition. First described by Alexander Tietze in 1921[1]. Tietze syndrome has been defined as a benign, painful, non-suppurative localized swelling of the costosternal, sternoclavicular or costochondral joints, most often involving the area of the second and third ribs. Only one area is usually involved and young adults are more commonly affected. [2] The syndrome is uncommon and self-limiting. Although the etiology of the disease is unknown, but preceding upper respiratory infections and excessive coughing have been described in some patients and it has been suggested that recurrent micro trauma of the anterior chest wall may be implied in the development of characteristic degenerative changes involving single or multiple upper costochondral junctions (usually 2nd and third costochondral junctions).

Although young adults are most frequently affected, the syndrome described by Tietze has been subsequently more commonly found in older people than previously reported and in this age group(i.e older age group) [3] it seems to have no sex or side prevalence. Because of the benign nature of this disease and its excellent prognosis, the treatment is usually symptomatic. The accumulated data stress the importance of a thorough clinical investigation and the need to exclude severe and life threatening conditions in the elderly. [4]

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Unlike costochondritis the markers of inflammation like C-reactive protein and ESR are increased in many cases and there is some debate to classify this entity as a seronegative arthropathy [2]

CLINICAL PRESENTATION

Tietze syndrome can present at any age but is most common in those aged 20 to 30 years. It is twice as frequent in men as in women (except in older age group in which the prevalence frequency is more even).[3]

Tietze'z syndrome usually comes on abruptly and is accompanied by localized swelling (and pain) and usually lasts for several weeks or months. It can appear as a complication of surgery on sternum, sometimes appearing months or years after the operation. The swelling varies from a slight firm enlargement sometimes seen in the early stages to a firm smooth or irregular mass obliterating the adjacent intercostal spaces. Tenderness varies from mild to extreme. It is most marked at the onset, tending to diminish greatly after the first week or fortnight, weeks or months before the swelling regresses. In the presence of a swelling, however, tenderness reappears with each fresh respiratory infection. In some cases there is additional pain and tenderness over an uninvolved rib near the spine.[5]. Occasionally however the onset is gradual.

So in summary two conditions need to be met if a diagnosis of Tietze syndrome is to be made with firmness:

(1) Painful and tender enlargement in the region of one or more of the costosternal junctions.

(2) This enlargement should not have been present previously, and should regress even without therapy.

The presentation of Tietze syndrome differs from costochondritis in that it usually affects typically only one joint whereas costochondritis may affect numerous costochondral joints [6]

The syndrome usually affects the upper ribs,

especially the second or third costochondral joints. (Unlike costochondritis which typically affects 2nd through 5th costal cartilages.). The pain is aggravated by physical activity, movement, coughing or sneezing. There is localized tenderness. The pain of Tietze syndrome can sometimes be quite severe and can mimic the pain of myocardial infarction. Although the pain usually disappears spontaneously, the swelling may persist long after the tenderness has disappeared.

DIAGNOSIS

It can be confused with other causes of chest pain, the pain may be confused with that of myocardial infarction and is usually unilateral on the left side.
Tietze syndrome and costochondritis are not completely synonymous, as Tietze syndrome is more localized and includes costochondral cartilage swelling, whereas costochondritis tends to be more diffuse and does not cause costochondral cartilage swelling.
Some of the earlier studies showed that although cartilage itself in that area is normal but the adjacent tissues show inflammatory infiltration[7], however later studies show that Tietze disease does have a recognizable pathological pattern which consists of an increased vascularity and degenerative changes with patchy loss of ground substance leading to a fibrillar appearance. Cleft formation may occur with mucoid debris which may undergo calcification. (Figure 1, 2&3) Hypertrophic changes may be present at the periphery.[8]
However it must be understood that a tissue like a cartilage can respond to a stimulus only in a limited no of ways so these pathological changes seen in tietze syndrome shed no light on the etiology of this disease.

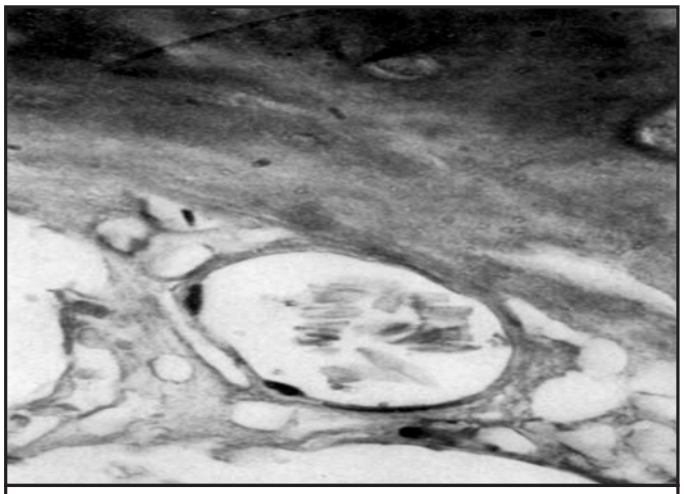

Figure 1: VACUOLAR STRUCTURES WITHIN THE HYALINE CARTILAGE

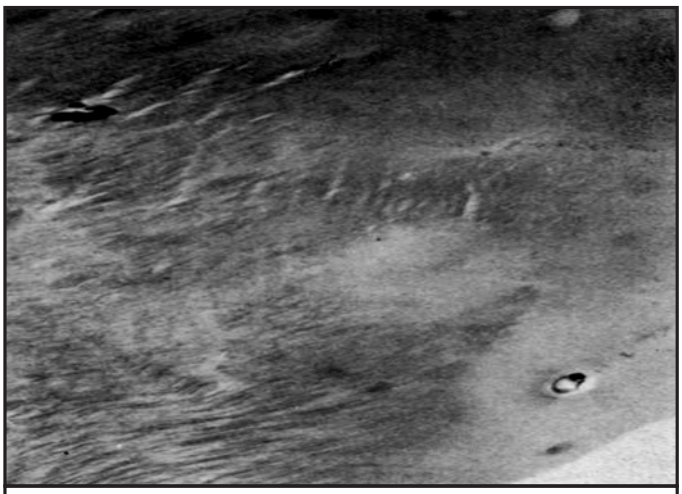

Figure 2: PATCHY AREAS OF UNMASKED COLLAGEN FIBRES ARE READILY SEEN (with some hint of increasd vascurality)

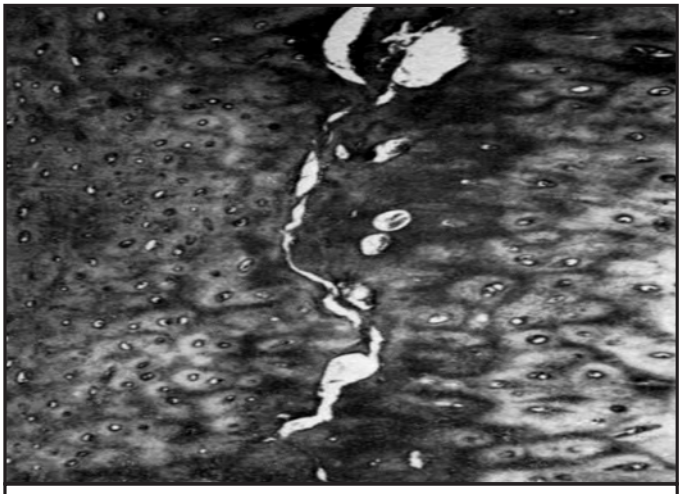

Figure 3: CLEFTS WITHIN THE CARTILAGE. THE WALLS OF THE CLEFTS ARE STAINED MORE DEEPLY AND THERE IS SOME MUCOID MATERIAL WITHIN THE CLEFTS.

(the histological pictures above are adopted from: H. U. Cameron and V. L. Fornasier .Tietze disease Clin Pathol. 1974 ; 27(12): 960–962.)
Diagnosis can usually be made by careful history and examination. Investigations may be required to rule out other possible causes of chest pain:[9]ECG to exclude cardiovascular conditions, chest X-ray to exclude other pathologies. (because the presence of chest tenderness may not help to distinguish between an ischaemic and non-ischaemic cause of chest pain). Although some studies have shown that the presence of chest wall tenderness has a positive correlation with normal myocardial perfusion.[10-18.] Ultrasound (and to a lesser extent CT scan) may have a role in assessment and diagnosis. [19]
Magnetic resonance imaging (MRI) may also be

useful and effective for some patients. The MRI pattern of primary Tietze syndrome was characterized as follows: Enlargement and thickening of cartilage at the site of complaint; focal or widespread increased signal intensities of affected cartilage on both TSE T2-weighted and STIR(Short TI Inversion Recovery is an inversion recovery pulse sequence with specific timing so as to suppress the signal from fat) or FAT SAT images(Fat suppression); bone marrow edema in the subcondral bone; vivid gadolinium uptake in the areas of thickened cartilage, in the subcondral bone marrow and/or in capsule and ligaments [20]
Magnetic resonance is an excellent technique to evidence both the cartilage and bone abnormalities; therefore it represents the elective method in the investigation of primary Tietze syndrome, due to its high sensitivity, diagnostic reliability and biological advantages thanks to the lack of ionizing radiation. [20]
Different imaging modalities are sometimes integrated to come to a final diagnosis as other seronegative arthropathies also come into the differencial diagnosis. [10]

MANAGEMENT

The patient can be reassured once the benign nature of the disease is confirmed. Rest, Non-steroidal anti-inflammatory drugs [21] and physical therapy can be useful. Ice packs applied to local swelling can sometimes help to reduce pain and inflammation. Local lidocaine analgesic patch (Lidoderm) application can also reduce pain. Local injection of long-acting corticosteroids may also be used. (Ultrasound is sometimes used to assess response to steroid injection)[19]. Intercostal nerve block may also help but is rarely required. Some research has shown that just a small amount of human calcitonin (a drug more commonly used to treat bone loss in postmenopausal women) improved Tietze syndrome patient's pain after a few days of treatment. According to Ricevuti, the hormone probably works because of the drug's benefit on the immune system and because of its pain-relieving effects. [22]

PROGNOSIS

The pain usually subsides within a few weeks, with some residual swelling persisting for longer periods of time. However, the course of the disease varies from spontaneous remission to persistent symptoms over years.

REFERENCES

1. Tietze A, Uber eine eigenartige, Haufung von Fallen mit, Dystrophie der Rippenknorpel, Berliner klinische Wochenschrift. 1921, 58: 29-831.
2. Aeschlimann A, Kahn MF. Clin Tietze syndrome: a critical review. Exp Rheumatol. 1990; 8(4):407-12.
3. Martino GCariati S, Elmore UPascarella , Corbelli G,C Hekmatdousttabrizi A, Carlomagno R. G Chir.1994 15(3):119-23.
4. M Ho, S Walker, F McGarry, S Pringle and T Pullar, Chest wall tenderness is unhelpful in the diagnosis of recurrent chest pain. QJM, 94 (2001), pp. 267–70.
5. A. K. Geddes, M D, Canad. M. A. J. Tietze Syndrome 1945,12 vol. 53i , 571-3
6. Karen Hudes, BSc, BS, DC, Can Chiropr. Low-tech rehabilitation and management of a 64 year old male patient with acute idiopathic onset of costochondritis. Assoc. 2008 ; 52(4): 224–8
7. J. LANDON and J. S. MALPAS Medical Division, Tietze Syndrome Ann Rheum Dis 1959; 18:249-54
8. H. U. Cameron and V. L. Fornasier .Tietze disease Clin Pathol. 1974 ; 27(12): 960–2.
9. Buntinx F, Knockaert D, Bruyninckx R, de Blaey N, Aerts M, Knottnerus JA, et al. Chest pain in general practice or in the hospital emergency department: is it the same Fam Pract. 2001; 8:586-9
10. Guglielmi G, Cascavilla A, Scalzo G, Salaffi F, Grassi W. Imaging of sternocostoclavicular joint in spondyloarthropaties and other rheumatic conditions .Clinical and experimental rheumatology. 2009 ;27(3):402-8.
11. Pasternak RC, Thibault GE, Savoiia M, et al. Chest pain with angiographically insignificant coronary arterial obstruction: clinical presentation and long term follow up. Am J Med 1982; 68:813–7.
12. Dart AM, Davies HA, Dalal J, et al. Angina and normal coronary arteriograms: a follow up study. Eur Heart J 1980; 1:97–100.
13. Wise CM, Semble EL, Dalton CB. Musculoskeletal chest wall syndromes in patients with non cardiac chest pain: a study of 100 patients. Arch Phys Med Rehabil 1992; 73:147–9.
14. Mukerji B, Mukerji V, Selukar R. The prevalence of rheumatological disorders in patients with chest pain and angiographically normal coronary arteries.Angiology 1995; 46:425–30.
15. Keavney B, Haider YM, McCance AJ, Skehan ID, Normal coronary angiograms: financial victory from the brink of clinical defeat? Heart 1996; 77:623–5.
16. Preman Kumarathurai, MDMohammed Kashaf Farooq, MDHenrik Wulff Christensen, MD,

DC, PhD, Werner Vach, PhDPoul F. Høilund-Carlsen, MD, DMSc, Muscular Tenderness in the Anterior Chest Wall in Patients With Stable Angina Pectoris is Associated With Normal Myocardial Perfusion. Journal of Manipulative and Physiological Therapeutics . 2008 Volume 31, Issue 5 , Pages 344-347,
17. Martino F, Ettorre GC, Macarini L, Guerrieri F, Tritto D. Diagnostic imaging of Tietze syndrome. Comparison of computerized tomography and ultrasonography Radiol Med. 1993 ;86(3):208-12.
18. Kemp HG, Vokanes PS, Cohn PE, et al. The anginal syndrome associated with normal coronary angiograms: report of a 6 year experience. Am J Med 1973;54:735–42.
19. Kamel M, Kotob H,.Ultrasonographic assessment of local steroid injection in Tietze syndrome, Br J Rheumatol. 1997.;36(5):547-50
20. Volterrani L Mazzei MA ,Giordano N ,Nuti R, Galeazzi M, Fioravanti.Source A. Magnetic resonance imaging in Tietze syndrome. Clin Exp Rheumatol. 2008 Sep-Oct; 26(5):848-53
21. E J Valtonen. Phenylbutazone in the treatment of Tietze syndrome Ann Rheum Dis 1967; 26(2): 133–135.
22. Ricevuti G. Effects of human calcitonin on pain in the treatment of Tietze syndrome, Clin Ther. 1985; 7(6):669-73.