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# **Tietze Syndrome**

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## **Continuing Education Activity**

Tietze syndrome is an atypical cause of anterior chest pain characterized by localized tenderness and non-suppurative swelling, usually associated with the 2nd or 3rd costal cartilages unilaterally. This condition is benign and self-limiting, with most patients experiencing complete relief of pain and swelling within weeks to months while utilizing conservative symptomatic treatment modalities. This activity reviews the evaluation and treatment of Tietze syndrome and highlights the role of the healthcare team in managing patients with this condition.

#### **Objectives:**

- Identify the etiology of Tietze syndrome.
- Describe the evaluation process for Tietze syndrome.
- Review the management options available for Tietze syndrome.
- Summarize interprofessional team strategies for improving care coordination and communication to advance the care of Tietze syndrome and improve outcomes.

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#### Introduction

Tietze syndrome (also called costochondral junction syndrome or chondropathia tuberosa), first described in 1921 by the German surgeon Alexander Tietze, is a rare and benign inflammatory condition characterized by chest pain and swelling at the costochondral junction.[1]

### **Etiology**

The exact etiology of Tietze syndrome is unclear. Some studies have postulated that multiple microtraumas to the anterior chest wall may trigger the development of Tietze syndrome. It may occur more frequently in certain conditions, such as psoriatic arthritis. Sometimes the development of the disorder may be preceded by chronic, excessive coughing, vomiting, trauma or impact to the chest, viral or bacterial infections, or surgery to the thoracic area.[2][3]

# **Epidemiology**

The precise prevalence and incidence of Tietze syndrome are unknown. Biological sex, ethnicity, geography, or occupation do not provide any insights into occurrence rates. Older youth and younger adults do seem to be affected more frequently than their older counterparts over the age of 40. Rarely are those at the far ends of the age spectrum affected, but cases have been reported.

## **Pathophysiology**

Tietze syndrome is described as a localized, painful, swollen, non-pustular mass commonly without rubor or erythema. It is most often associated with the cartilage of ribs two or three and contained to one side in approximately 70% of patients, though it has been identified at the sternoclavicular and xiphisternal joints.[6]

There have been discussions regarding the possibility that this disease process is inflammatory and may be part of a more comprehensive seronegative pathology.[3] The symptoms are often self-limiting, but Tietze syndrome recurrences are possible.

## Histopathology

The costal cartilage histopathology is notable for increased vascularity and hypertrophy of the peripheral cartilage, which indicates that there is proliferation occurring. The perichondrium appears to be unaffected. Other findings include mucopolysaccharide containing clefts that may undergo calcification.[7]

## **History and Physical**

Patients typically present complaining of acute chest pain with no history of trauma. Due to the younger population in which Tietze syndrome tends to occur, it is not uncommon for the patients to be otherwise healthy. The pain is sharp and stabbing over the swollen area and can radiate to the shoulder and proximal arm. Heat and erythema are not usually associated with the swelling. The patient's discomfort can be made worse with the movement of the ipsilateral arm, movement of the torso, coughing, sneezing, and deep breathing.[2]

Cardiac, pulmonary, and neurological exams are typically benign in connection with Tietze syndrome, and any underlying cardiopulmonary problems discovered upon exam should receive further attention

#### **Evaluation**

Tietze syndrome is a diagnosis of exclusion after a thorough workup of life-threatening or more common diseases has been completed. An electrocardiogram (ECG) should be performed on all patients that present with acute chest pain. Lab results for this syndrome are non-specific. Biopsy of the costal cartilage can lead to a more prompt diagnosis if obtained early in the disease progression. Ultrasound has proven to be the most effective modality to assist in the diagnosis of Tietze syndrome as it can quickly demonstrate soft tissue swelling at the site of inflammation. Another useful diagnostic tool, though not specific, is nuclear magnetic resonance (NMR), which accurately identifies alterations in the neighboring fat tissue and bone marrow due to inflammation. These inflammatory changes can lead to compression and joint narrowing of the affected articulation.[2] Laboratory work up may also show elevated inflammatory markers such as ESR or CRP.

Radiographs are typically read as normal, while computed tomography (CT) may show a slight focal swelling or mild sclerosing of the symptomatic joint.[8] In some more recent case reports, positron emission tomography CT-fluorodeoxyglucose (PET/CT-FDG) was obtained, which

allowed for the visualization of hypermetabolic activity at the symptomatic joint with dense calcification.[9][10] This type of imaging, though not used frequently at this time, could, in the future, become a regular tool in the workup of Tietze syndrome. In summary, imaging and labs should be obtained when a patient presents with acute chest pain, but in Tietze syndrome, the results are best utilized as a method of ruling out other potential causes.

## **Treatment / Management**

The keystone of Tietze syndrome treatment is conservative therapy and reassurance that this disease process will often resolve on its own with no permanent sequelae, many times within a few weeks. There is, however, the potential for it to last months, or even up to a year. First-line medical management is oral or topical anti-inflammatory and analgesic agents.

If those do not provide significant relief, patients may benefit from a focused injection of local anesthetic, steroid, or both at the site of maximum swelling, which should be visualized via sonogram. Some patients have reported benefits with warming pads applied to the affected area. There are reports in which cartilage has been resected as a treatment option in persistent, severe cases. However, this is not generally recommended and should only be considered on a case by case basis.[2]

## **Differential Diagnosis**

As a diagnosis of exclusion, it is vital upon initial presentation of acute chest pain that a wide differential diagnosis is considered. Tietze syndrome is most commonly misdiagnosed as costochondritis, as the latter also presents as anterior chest wall pain that is reproduced with palpation at the sternochondral and costochondral junctions. Costochondritis, however, is associated with multiple ribs typically ranging from 2 to 5 and is not associated with localized swelling over the affected joints. Ultrasound will not demonstrate significant findings with costochondritis, whereas sonogram is a mainstay in aiding a Tietze syndrome diagnosis.

Other pathologies to rule out include acute coronary syndrome, hypertensive crisis, inflammatory processes or infections of the lungs and the accompanying pleura, malignancies, trauma associated fractures in the chest, rheumatoid or pyogenic arthritis, gastroesophageal reflux disease, or psychogenic disorders.[2][5]

# **Prognosis**

Tietze syndrome is a self-limiting disease with a good prognosis. Most patients report complete resolution of symptoms within 1 to 2 weeks with conservative management, but there have been reports of the characteristic swelling lasting for up to one year. Reoccurrences are also possible.

# Complications

There are usually no complications directly associated with Tietze syndrome, other than those potentially as a result of injections, medications, or surgery. These may include infection, medication reactions, medication side effects, and scarring.

#### **Deterrence and Patient Education**

Patients should be advised to avoid strenuous activity for 1 to 2 weeks, as it is likely to

exacerbate their discomfort and possibly delay the resolution of their symptoms. Providers should follow these patients closely if there is a concern for worsening symptoms or increasing swelling.

## **Enhancing Healthcare Team Outcomes**

The most important point regarding Tietze syndrome is that it is a diagnosis of exclusion. In and of itself, it is not a deadly disease. To decrease the patient's physical discomfort, mental anguish, wasted time, and expenses, practitioners should be aware of the clinical presentation, identify the condition, complete a thorough workup for any acute life-threatening disorders that may be possible, and inform patients about this condition to reassure the patient about this generally benign and self-limited condition.[5] Communication between the interprofessional care team is a key element in making an appropriate and timely diagnosis. When other possibilities have been ruled out, reassurance should be provided. The patient can then usually be managed by their primary care team on an outpatient basis as needed.

#### **Review Questions**

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