

Avascular necrosis of the sternal end of the clavicle:

A case report

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Avascular necrosis of the sternal end of the clavicle was described by Friedrich³ in 1924 and is known as Friedrich disease. Since then, few cases have been reported in the literature. Most patients have been women and without histologic confirmation of the disease. The disease is rare, and the etiology is unknown. In general, it presents with pain in the sternoclavicular joint (SCJ) and an increase in the volume of the sternal end of the clavicle that progresses to an insidious functional limitation of the shoulder joint.⁵ Conservative treatment results in remission of the symptoms in most patients, and resection of the sternal end of the clavicle is reserved for resistant cases.⁹ We report a man with Friedrich disease confirmed by histologic examination who underwent resection of the sternal end of the clavicle.

CASE REPORT

A 42-year-old man complained of increased volume and pain in the right SCJ of 1-year duration. There was no history of trauma, infection, steroid treatment, or other apparent cause. The result of a biopsy 6 months earlier in the anterior region of the SCJ was inconclusive. The pathology report revealed the presence of bone and mature cartilaginous tissue without signs of malignancy. At the time, conservative treatment was instituted with physiotherapy, analgesics, and nonsteroidal anti-inflammatory medications.

When the patient arrived on our service, he had a 1.5-cm scar in the anterior region of the SCJ, without signs of inflammation. He also had a solid, painful, palpable mass of about 2.0 cm in the sternal end of the clavicle (Figure 1). Motion of the shoulder showed painful limitation of about 10° at the end of the elevation, without change in internal or external rotation. Complete blood cell count, hemoglobin level, and sedimentation rate were within normal limits. Radiology and computed tomography (CT) scan of the SCJ demon-

strated the undefined outlines and areas of bone destruction mainly in the inferior aspect of the sternal end of the clavicle (Figures 2 and 3).

Initially, the patient was treated conservatively with non-steroidal anti-inflammatory drugs, analgesics, and physiotherapy. After 6 months without improvement, he underwent surgery with resection of the sternal end of the clavicle and pathologic examination of the resected bone segment. A curvilinear incision of about 6.0 cm in length was made over the anterior region of the SCJ. The anterior sternoclavicular capsule was identified and opened for access to the sternal end of the clavicle. The costoclavicular ligament was identified after careful subperiosteal dissection, and the portion of the clavicle medial to the ligament was resected.

The resected specimen showed an osteophyte at the inferior portion of the joint surface and partial destruction of the cartilage. Microscopic examination of the resected specimen demonstrated small areas of secondary arthrosis with thickening of the bone trabeculae; however, large necrotic areas predominated, characterized by empty bone lacunae of the Haversian system, confirming the diagnosis of Friedrich disease (Figure 4). At the 6-month postoperative follow-up, the patient was free of pain, with normal motion and complete remission of the deformity at the SCJ.

DISCUSSION

Friedrich disease, characterized by avascular necrosis of the sternal end of the clavicle, is rare and its characteristics are similar to those of Perthes, Kohler, Kienbock, and Preiser diseases.³ A survey of the literature found few descriptions, and they mostly discuss the clinical and radiographic characteristics of the disease.^{1,2,5,7}

The etiology of the avascular necrosis of the sternal end of the clavicle is unknown; nevertheless, there are some theories on probable causes. Vandor et al¹⁰ link the disease to a lesion of the accessory spinal nerve that occurs during surgeries that involve extensive neck dissection. Friedrich³ and Knestch⁶ support a theory of repetitive microtrauma. Willert et al⁵ suggest that a congenital malformation of the sternal end of the clavicle may be the cause. We failed to find any cause for necrosis of the sternal end of the clavicle in this case.

The disease affects female patients with greater frequency and in ages ranging between 6 and 58 years, with few cases described in men.⁷ It is usually unilateral, with only 1 report of a bilateral case.⁹ Clinically, the patients present with an increase in volume and pain in the SCJ, without a history of trauma.

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J Shoulder Elbow Surg 2008;17:e31-e33

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1058-2746/2008/\$34.00

doi:10.1016/j.jse.2007.08.008



Figure 1 Increased volume in the anterior region of the right sternoclavicular joint.

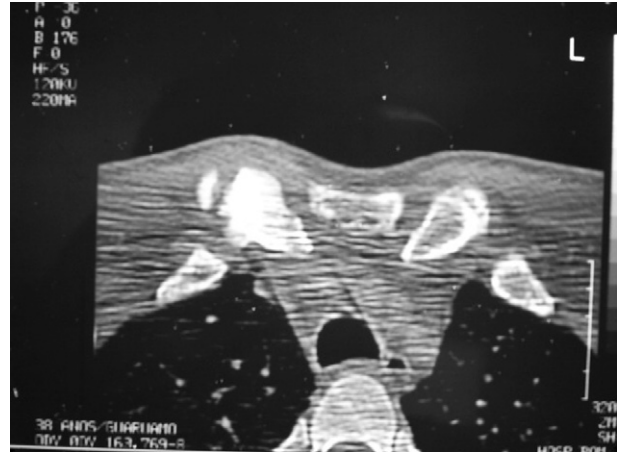


Figure 3 Computed tomography scan shows bone destruction of the medial clavicle.

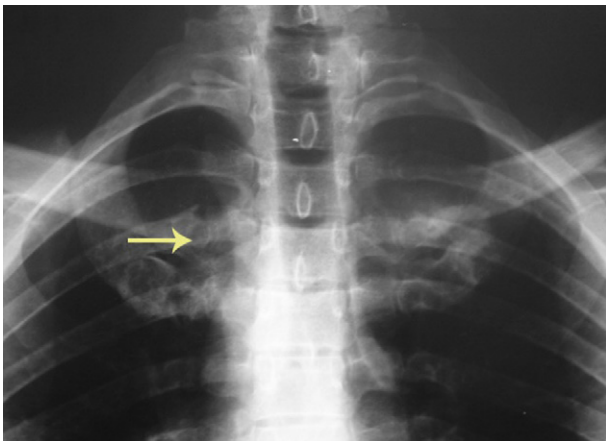


Figure 2 Radiograph shows erosion and bone defect of the sternal end of the right clavicle (arrow).

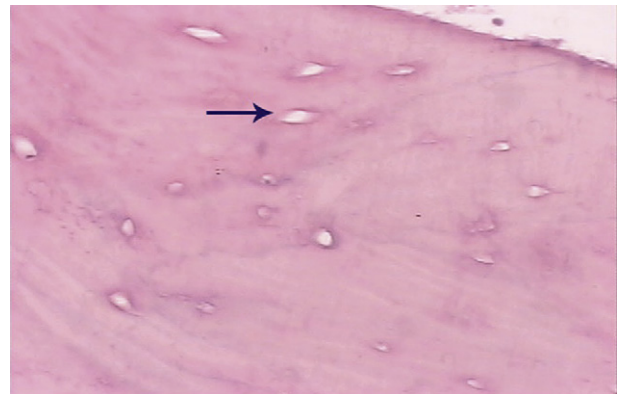


Figure 4 Photomicrograph demonstrates avascular necrosis of the sternal end of the clavicle (empty bone lacunae; arrow).

The SCJ is a synovial joint with movement in all planes including rotation. During normal movement of the shoulder, the SCJ can elevate from 30° to 35°, flex and extend 35°, and rotate on its axis from 45° to 50°.⁸ Changes in articular biomechanics in more advanced stages of the disease can cause functional limitation of the shoulder. This was seen in our patient, who showed painful limitation of 10° at the end of shoulder elevation. The patient usually endures the symptoms for months before seeking medical assistance.

The diagnosis is generally arrived at late. Owing to the rarity of Friedrich disease, it is not considered during the initial diagnostic investigation. This was the case in our patient, who sought medical help only a year after the onset of symptoms and had conservative treatment for a long period without a definitive diagnosis. Avascular necrosis of the sternal end of the clavicle should be differentiated from osteoarthritis, neoplasia, tuberculosis, osteomyelitis, and subluxation of the SCJ.¹

Radiography reveals the presence of irregularity of the SCJ and bony defect in the sternal end of the clavicle. These

changes are more pronounced on the CT scan.⁹ The radiograph examination of this joint, not only for necrosis but for other diseases as well, is not very sensitive or specific because of the overlapping of other bony structures. The pathologic examination generally reveals cystic areas filled with necrotic tissue characterized by empty bone lacunae of the Haversian system surrounded by intact bone.⁵

The treatment of Friedrich disease is controversial and ranges from conservative management to surgical resection of the medial end of the clavicle. Those who favor surgical treatment claim that the long-term results of conservative treatment are not encouraging.^{5,7} The proponents of conservative treatment report that most patients have complete remission of symptoms and that surgery should be performed only in rare cases that fail to improve with conservative measures.^{7,8} Some authors believe that this disease is self-limited and progresses to spontaneous remission.¹⁰ Heinemeier et al⁴ are of the opinion that the disease has a foreseeable outcome that can be divided into different stages. We were unable to confirm this type of outcome,

either in our patient or in the literature. The few reports in the literature mention that the initial treatment should be conservative and that surgery is rarely indicated. Our opinion is that surgery is an effective treatment for Friedrich disease mainly in cases that progress to significant pain and limitation of the rotation arc of the shoulder joint.

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