
症 例

Friedrich's Disease: A Case Report

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Received for Publication, Feb. 15, 1991.

Summary

An eight-year-old girl was presented with painful swelling of the sternoclavicular joint. Radiological and scintigraphic examinations lead to a diagnosis of Friedrich's disease, aseptic necrosis of the sternal end of the clavicle. It is a rare condition which is to be differentiated from osteomyelitis, arthritis, or tumor. Because it is a self-resolving, benign condition, awareness of this disease will save the patient from unnecessary surgery.

Introduction

Friedrich's disease is the avascular necrosis of the sternal end of the clavicle that follows a benign course. Only 24 cases have been described in the literatures since the first report by FRIEDRICH⁽⁶⁾ in 1924. The purpose of this communication was to present the clinical and radiological findings and benign nature of this condition seen in a Japanese girl diagnosed as having Friedrich's disease. The diagnostic value of the bone scintigraphy in this case is also described.

Case report

An eight-year-old girl was presented in our clinic with pain and swelling in the region of her left sternoclavicular joint for one month. She had no fever or history of trauma. The pain increased on trying to move her left shoulder. On physical examination a tender swelling measuring two by three centimeters could be palpated at her left sternoclavicular joint, and there was an anteroposterior instability of the sternal end of the clavicle on deep palpation (Fig. 1). Anteroposterior roentgenogram revealed a bony destruction in the left clavicle at the sternal end. The bone lesion was more easily visible by tomography, which disclosed irregular bone resorption and sclerosis in the medial clavicular end (Fig. 2)

Key words: Friedrich's Disease, Clavicle, Osteonecrosis

索引語: Friedrich 病, 鎖骨, 骨壊死.

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Abbreviation: VDRL, Venereal Disease Research Laboratories. CRP, C-reactive protein. Tc-MDP, Technetium methylene diphosphonate.

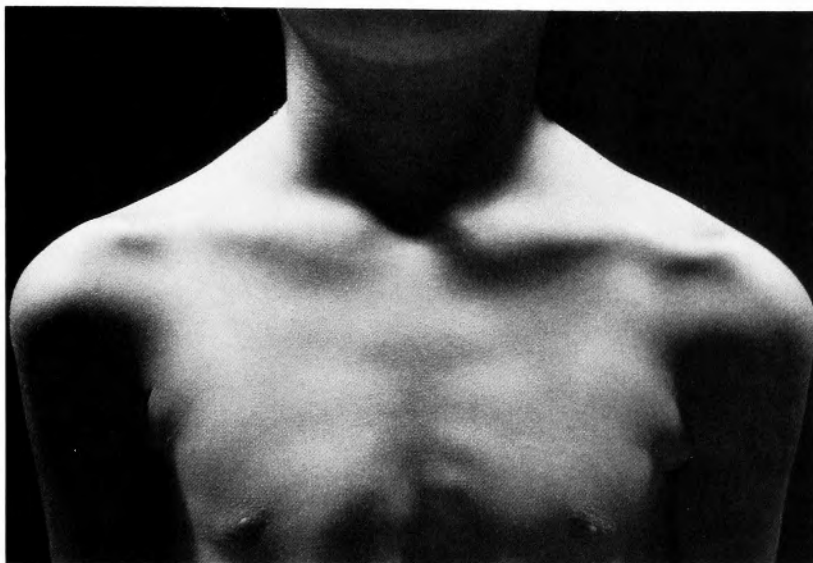


Fig. 1. Clinical appearance of the patient. Painful swelling over the left sternoclavicular joint was noted.

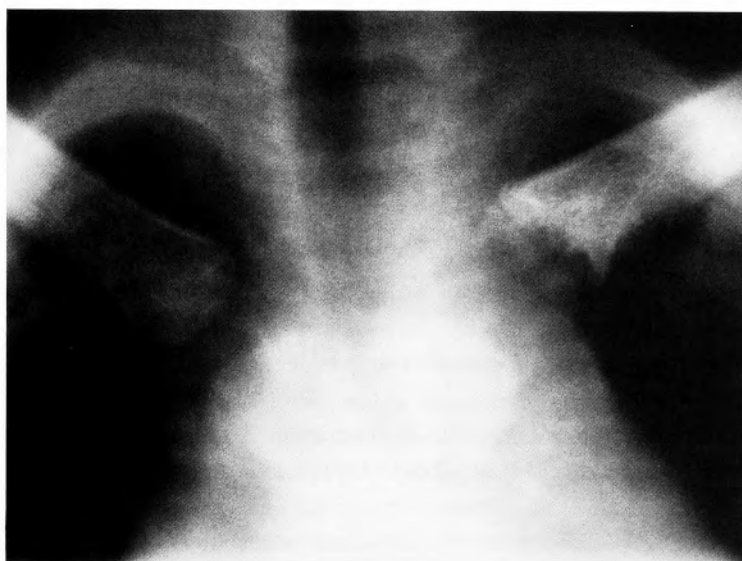


Fig. 2. Tomogram of the clavicles showing bony destruction with resorption and sclerosis at the sternal end of the clavicle.

Erythrocyte sedimentation rate was 6 and 16 millimeters in the first and second hour, respectively. the white cell count, haemoglobin and haematocrit were normal. Serum alkaline phosphatase level was normal. Latex and Rose-Waaler tests were negative, and so was the antistreptolysin titre. the VDRL test was nonreactive. Liver and kidney function tests were also normal and CRP was negative.

Bone scintigraphy performed 10 days after the first visit using ^{99m}Tc -MDP showed increased up-

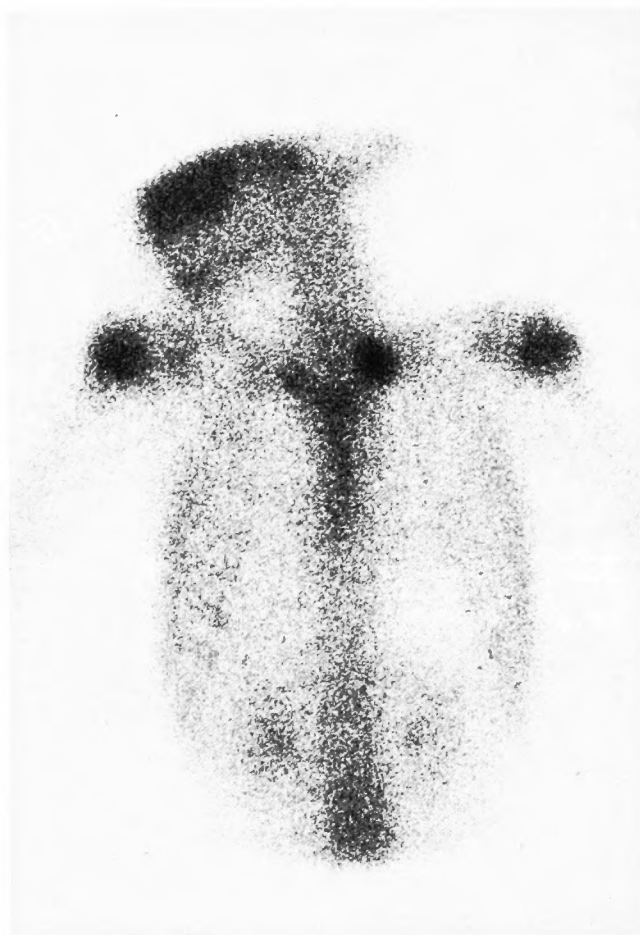


Fig. 3. ^{99m}Tc -MDP bone scintigram showing an increased uptake in the left sternoclavicular joint.

take in the left sternoclavicular joint (Fig. 3). A diagnosis of aseptic necrosis of the sternal end of the clavicle was made, and it was decided she should only be observed because there was no sign of inflammation.

The unstable sternoclavicular joint was stabilized, and pain was relieved by application of a figure-of-eight clavicle band worn throughout the period of six months of observation. At ten months after the onset of the symptoms, subluxation of the sternoclavicular joint had disappeared; tomography showed regeneration of the clavicular end (Fig. 4), and no marked uptake by bone scintigraphy was observed.

Discussion

Friedrich's disease is a rare condition which was first described by FRIEDRICH in 1924⁶⁾. Including FRIEDRICH's two original cases, there has been only twenty four cases in the literatures from western countries^{1,4,5,7,8,9,11,12)}. The age of the patients range from 6 to 58 years and there is no di-

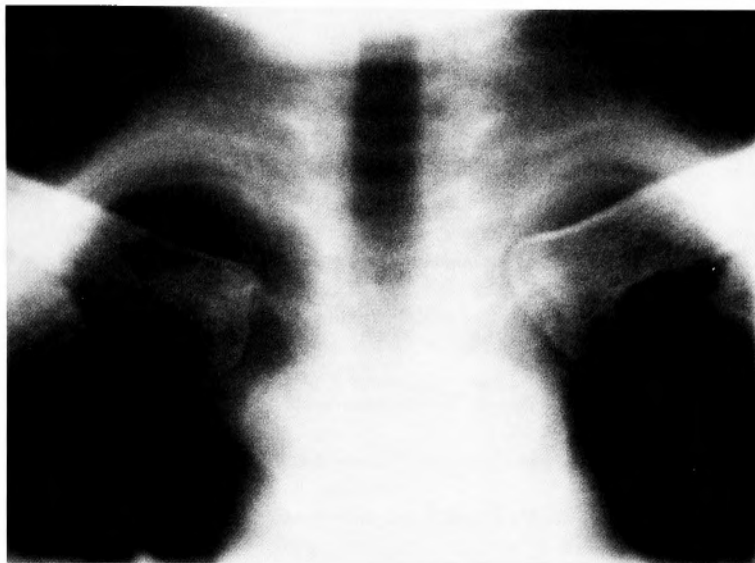


Fig. 4. Tomogram of the clavicles ten months after the onset of symptoms showing regeneration of the clavicular end.

ference in incidence between sexes.

Many conditions affect the sternoclavicular joint¹³⁾ and Friedrich's disease may be confused by congenital anomaly, arthritis, osteomyelitis, tumor or condensing osteitis²⁾. Awareness of this rare disease and careful roentgenographic assessment especially by tomography will lead to a correct diagnosis.

In the early description of this disease, some cases were operated for biopsy. HEINEMEIER and TORKLUS even suggested resection of the medial clavicular end in a case of marked reduced shoulder movement⁹⁾. LEVY et al.¹¹⁾ in their report of four cases described that biopsy is an unnecessary procedure for such a benign disease. In the present case we applied bone scintigraphy using ^{99m}Tc-MDP which has not been described in the literature concerning this disease. There was a high uptake of nuclide in the sternoclavicular joint, which is a consistent finding generally seen at the reparative stage of osteonecrosis¹⁰⁾. Although the etiology of this disease is unknown, it is thought to be due to aseptic necrosis of the sternal end of the clavicle, as was suggested by FRIEDRICH in 1924.

Our case was presented with painful anterior subluxation of the sternoclavicular joint which responded well to a figure-of-eight clavicular bandage. This type of immobilization is recommended in traumatic sternoclavicular joint subluxation²⁾. With this immobilization the patient followed a benign self-resolving course with little discomfort. Awareness of this benign disease will spare unnecessary surgery including biopsy.

Acknowledgment

Gratitude is extended to Mr. K. Wake and Ms. M. Sato, Department of Medical Photography, Kokura Memorial Hospital for preparation of the figures.

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和文抄録

Friedrich 病 の 1 例

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胸鎖関節に、痛みをともなう腫脹を訴えて来診した、8歳、女性に対し X 線、骨シンチ所見などより、鎖骨の内側端におこる骨壊死、Friedrich 病と診断した。保存的治療により症状は軽快し、10ヵ月後の X 線で

治癒を確認した。本症はまれな疾患で、骨髓炎、関節炎、腫瘍との鑑別が必要である。良性の自然に緩解する疾患であるので、本疾患を識別し、不要な手術や生検をさけることが重要である。