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POLYOSTOTIC FIBROUS DYSPLASIA

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Key-word: Bones, fibrous dysplasia

Background: A 51-year-old male was referred to the hospital with pain on the right side of the thorax, without any apparent trauma. There was no medical history. He had a nonproductive cough and smoked 15 cigarettes a day. The patient did not have fever or weight loss and was not feeling ill. There was no history of asbestos contact.
Laboratory results showed no signs of infection or liver disease.

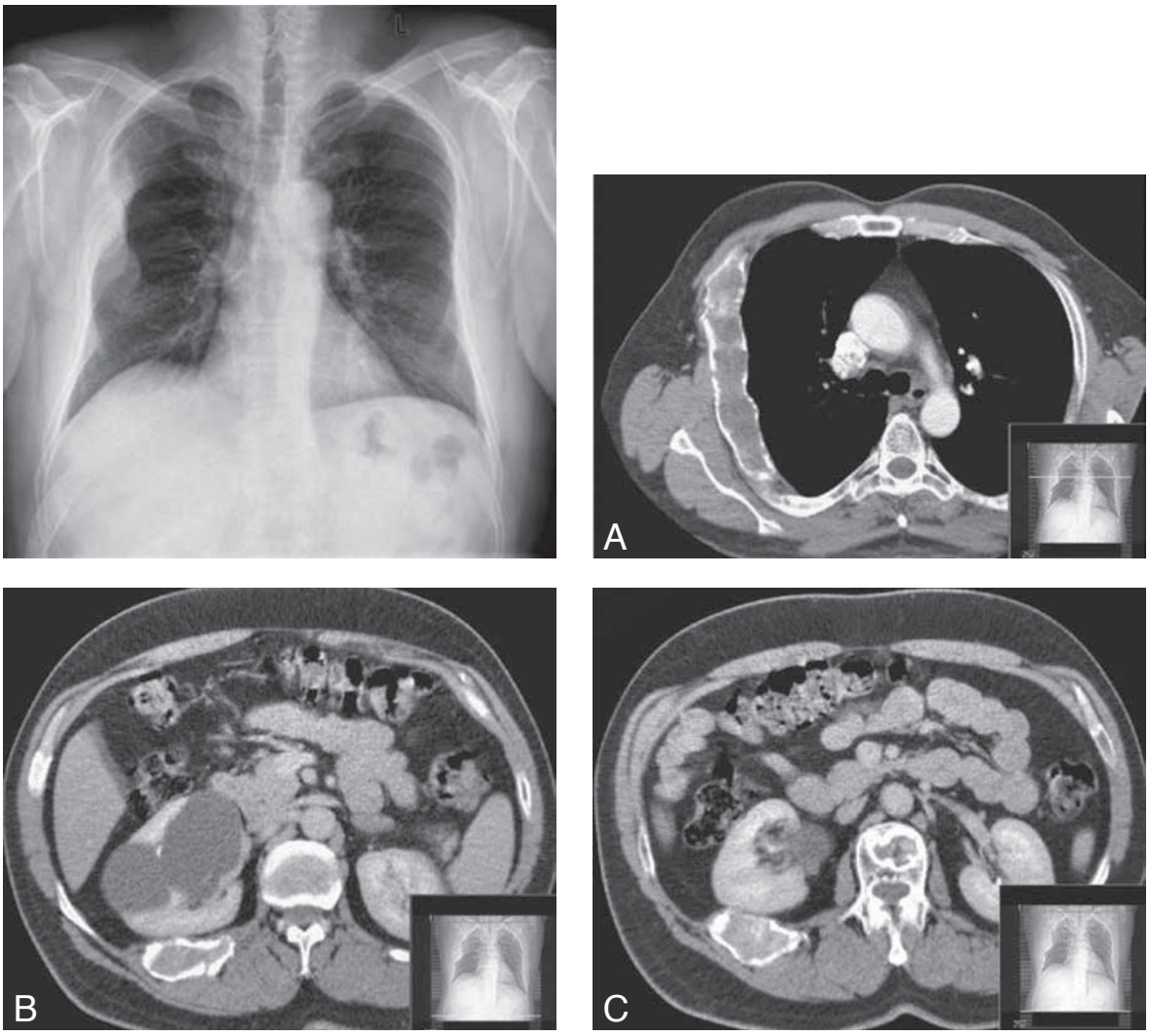

Fig. 1 | 2A / 2B | 2C

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Work-up

On chest radiography (PA-view) (Fig. 1), an enlarged, ground-glass aspect of the entire 4th right rib is seen, as well as peripheral wall thickening at the level of the right upper lobe.

On contrast-enhanced CT scan of the thorax and upper abdomen (soft tissue window setting) (Fig. 2), an enlarged 4th right rib is seen (A), with peripheral zone of sclerosis and no internal matrix formation. There is a fracture of the lateral side of the 4th rib. There are similar findings in the 12th right rib on the dorsal side (B), with a fracture on the dorsal side. Large cortical kidney cysts are visible on the right side. A morphologically similar lytic lesion with a sclerotic edge is seen in the body of the 2nd lumbar vertebra (C).

Radiological diagnosis

Based on the image findings the diagnosis *polyostotic fibrous dysplasia* was made.

The pain was most likely caused by the rib fractures. For a complete overview of the locations of the lesions, further imaging by means of a PET-CT is planned.

Discussion

Fibrous dysplasia (FD) is a sporadic benign skeletal disorder that can affect one bone (monostotic form) or multiple bones (polyostotic form). The latter may form part of the McCune-Albright syndrome (MAS) or of the Jaffe-Lichtenstein syndrome (JLS). JLS is characterized by polyostotic FD and café-au-lait pigmented skin lesions, while MAS has the additional features of hyperfunctional endocrinopathies manifesting as precocious puberty, hyperthyroidism or acromegaly. Patients with polyostotic FD often have renal phosphate wasting. The disease, however, has a wide clinical spectrum, so many patients are asymptomatic. Diagnosis relies on radiographs and pathology.

There is no gender prevalence for FD. The monostotic form is more common and affects the 20-30 years age group. Polyostotic FD has its onset mainly in children younger than 10 years of age. The lesions grow with the child, stabilize after puberty, and most commonly involve craniofacial bones, ribs, and metaphysis or diaphysis of the proximal femur or tibia. The occurrence ratio of polyostotic to monostotic FD is 3:7.

Signs and symptoms of FD include bone pain, pathological fractures and bone deformities. Serum alkaline phosphatase is occasionally elevated, but calcium, parathyroid hormone, 25-hydroxyvitamin D and 1,25-dihydroxyvitamin D levels are normal in most cases of FD. Persons with extensive poly-^ostotic FD may have hypophosphatemia, hyperphosphaturia and osteomalacy. Malignant transformation is rare and is usually precipitated by radiation therapy.

Bibliography

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