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Pediatric Radiology Quiz – Case 17

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A 4-year-old child presented to the Outpatient Pneumonology Unit of our hospital due to cough and low grade fever. Clinical examination revealed café-au-lait spots. The child had undergone a chest X-ray the previous week, without any reported significant findings. Second reading of the films revealed sclerosis and thinning of right lower thoracic ribs, along with widening of the intervertebral foramina (fig. 1). Further physical examination revealed soft tissue masses at the right posterior hemithorax. Subsequent focused soft tissue ultrasound examination of the thorax revealed in the right lower posterior interspaces, multiple, well circumscribed hypoechoic lesions, without detected vascularity (figures 2–3). These lesions were slightly palpable and relatively soft and they had intact contact with the respective posterior parts of thoracic ribs, causing a smooth scalloping on their cortical surface. Clinical findings along with the radiological findings were suggestive of the final diagnosis.

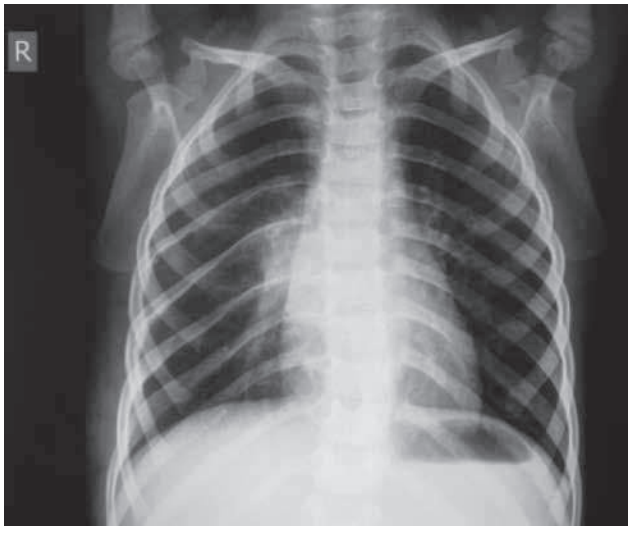

Figure 1. Chest X-ray, that reveals thinning and sclerosis of posterior part of 6th to 8th right thoracic ribs, along with widening of their intervertebral foramina.

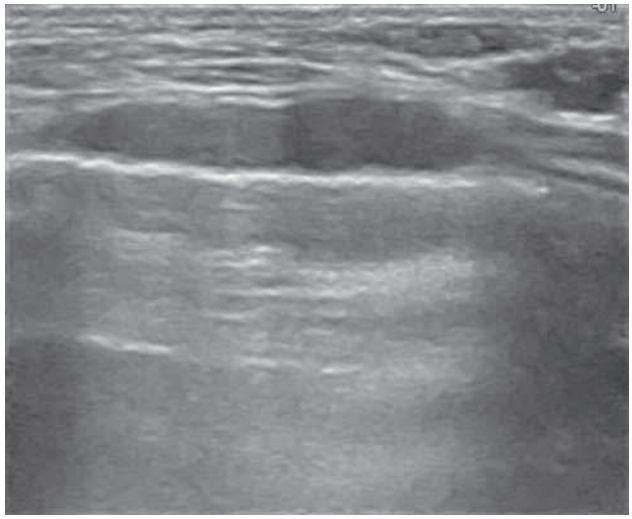

Figure 2. Ultrasound image in parallel with a posterior right intercostal space. A soft tissue, hypoechoic mass with well-defined margins, that causes smooth scalloping to the affected ribs.

Comment

Neurofibromatosis (NF) is an autosomal dominant disease. Several types of NF have been described, and type 1 (NF1), also denominated peripheral or classic NF, is the most common type. Its main clinical presentations include café-au-lait spots, cutaneous neurofibromas, axillary and or inguinal ephelides or freckling, plexiform neurofibromas and Lisch nodules.

The commonest bone abnormalities observed in NF1 patients include scoliosis, kyphoscoliosis, erosion of the anterior or posterior wall of vertebral bodies, intervertebral foramina widening, growth disorders, pseudoarthrosis, long bone cortical thinning, bone cysts, costal arch erosion, pedicle thinning, dysplasia of sphenoid wing, cranial osteolytic lesions, facial/mandibular deformities, subperiosteal bone proliferation, bone compression due to soft-tissue tumors (neurofibromas, dura mater dysplasias, intrathoracic meningocele). Thoracic neurofibromas are benign tumors involving the nerve roots

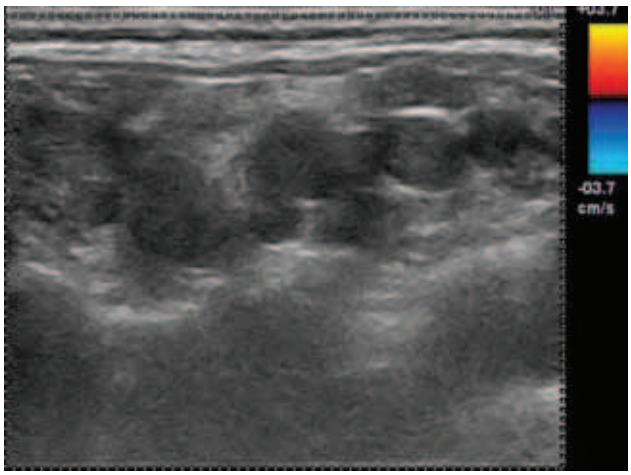

Figure 3. Color Doppler US image, perpendicular to the axis of posterior intercostal spaces. Multiple, well-defined, soft tissue masses that lack vascularity, observed in different levels.

of the spinal cord unilaterally or bilaterally, which usually need further imaging investigation with computed tomography (CT) or magnetic resonance imaging (MRI).

References

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