Chest: far beyond heart and lungs.

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Learning objectives

Demonstrate the imaging findings in syndromes involving the chest wall, like SAPHO, Poland Syndrome, Tuberous sclerosis (ET), Melorheostosis and Hereditary multiple exostoses (HME).

Review the contribution of Multidetector CT (MDCT) in the management and diagnosis of these syndromes.
Background

**SAPHO Syndrome.** This syndrome was described in 1987 as an association of rheumatologic and cutaneous features. The acronym SAPHO refers to synovitis, acne, palmoplantar pustulosis, hyperostosis and osteitis. However, not all the syndrome components need to be present for the diagnosis, particularly the dermatologic components, as osteoarticular involvement can manifest alone without any skin lesion. The aseptic skeletal inflammatory process is the most frequent manifestation and generally affects the sternoclavicular joints (70-90%). This finding is known as sternoclavicular hyperostosis and can be accompanied of pain and soft tissue swelling. SAPHO syndrome is observed mainly in young or middle-aged adults but can affect all ages and occurs with equal frequency in men and women. Differential diagnosis includes osteomyelitis, osteosarcoma, Ewing's Sarcoma, Paget's disease, metastasis, and avascular necrosis of bone.

**Poland Syndrome.** This is an uncommon congenital condition of the chest wall. It is an autosomal recessive condition, characterized by partial or total absence of the greater pectoral muscle and ipsilateral syndactyly. Associated conditions include bony dysostoses affecting the ipsilateral hand, pectus excavatum, anomalies of the upper limb, absence of the smaller pectoral muscle, hypoplasia of latissimus dorsi and serratus anterior, hypoplasia or aplasia of nipple and breast, lung herniation and hypoplasia of the ipsilateral second to fifth ribs. Bilateral involvement is rare. It is more frequent in males than females (3:1) and involves the right side in 75% of patients. Increased incidences of lung cancer, breast cancer, non-Hodgkin's lymphoma, and leukemia have been reported in these patients.

**Melorheostosis.** This is a rare sclerosing bone dysplasia, also known as Leri disease. It's characterized as flowing hyperostosis (Candle Wax Sign), and combines endochondral and intramembranous ossification failure. It doesn't show any evidence of genetic inheritance. The age of the presentation varies, but it usually remains occult until late adolescence or early adulthood. The disease usually follows a chronic progressive course, occasionally leading to substantial disability that may result in amputation. It may manifests as monostotic (single bone), monomelic (single limb) or polyostotic (multiple bones). There are reports of several associated clinical entities, including soft tissue fibrosis, soft tissue periarticular ossification, vascular tumors and malformations (hemangiomas, glomus tumors, and arteriovenous malformations), which suggest an association with a defect in angiogenesis.

**Tuberous Sclerosis (TS).** This is an autosomal-dominant disorder with incomplete penetrance. It can affect both sexes and all ethnic groups. A classical triad of epilepsy, mental and adenoma sebaceum is described but is uncommonly seen at
clinical examination; therefore radiologic examinations can play an important role in the diagnosis of TS. Abnormal radiologic findings include central nervous system (CNS) lesions such as cortical hamartomas, white-matter abnormalities, and subependymal nodules (hamartomas). Patients may also have subependymal giant cell astrocytomas. These intracranial neoplasms can result in obstructive hydrocephalus. Retinal lesions, nonatheromatous vascular stenoses, aneurysms, and mild ventricular enlargement without astrocytomas can be present. TS is a multisystemic disorder and common findings include hypopigmented skin lesion, periungueal fibromas, angiomyolipomas, cardiac rhabdomyoma, lung cists and micronodules and focal sclerotic bone lesions. The presence of pulmonary lymphangioleiomyomatosis (LAM), multifocal micronodular pneumocyte hyperplasia (MMPH), multiple renal cysts or angiomyolipomas and four or more focal bones sclerotic lesions suggest tuberous sclerosis. LAM affects most exclusively middle-aged women and is characterized by idiosyncratic smooth muscle cell proliferation (LAM cells) which leads to lung cysts, systemic lymphatic abnormalities and tumors. MMPH is a hamartomatous proliferation of the pneumocyte type II cell along the alveolar septa that exhibits multiple micronodules (1-8 mm) with random distribution.

**Hereditary multiple exostoses (HME).** This condition is classified by some authorities in the category of bones dysplasias. It is a hereditary, autosomal-dominant disorder with incomplete penetrance in females. Approximately two thirds of affected individuals have a positive family history. Males are affected two times more than females. The knees, ankles and shoulders are the sites most frequently affected by this disease. The osteochondroma is a lesion characterized by a cartilage-capped bony projection on external surface of a bone. This lesion usually stops growing at skeletal maturity. It is the most common benign bone lesion, comprising approximately 20% to 50% of all benign bone tumors, and is frequently diagnosed in patients before third decade.
MDCT can provide us an enormous amount of information. In a single chest scan with volumetric acquisition it is possible to use multiplanar reformations (MPR) or Volume Rendering post-processing to easily interpret all the data.

**SAPHO Syndrome.** Radiographic findings typically include hyperostosis and osteosclerosis with narrowing or ankylosis of the joint space. Destructive changes, such as osteolytic areas or erosions, also may be noticed. The most frequent site of aggression is the sternoclavicular joints (70-90% - see Figs. 1-3), but less frequently, involvement of spine, sacroiliac joint or long tubular bones may occur. Radionuclide bone scanning may show increased uptake in asymptomatic regions.

**Poland Syndrome.** Chest radiography usually shows unilateral hyperlucency with absence of the normal axillary fold on the affected side that mimics a radical mastectomy (Figs. 4-5). Rib deformities are found occasionally (Fig. 6). Diagnosis may always be made on MDCT or MRI, both of which will show absence or hypoplasia of pectoral girdle musculature (Fig. 5, 8-9). Also absence of the sternocostal head of the greater pectoral muscle, hypoplasia or aplasia of nipple, breast (Fig. 5) and serratus anterior muscle (Fig. 7) is always visible with these techniques.

**Melorheostosis** has rarely been reported in the axial skeleton. Although it can also be seen in the ribs, hands and feet, it predominantly affects the appendicular skeleton and is most common in the long bones of the upper and lower extremities. The classical presentation is a flowing hyperostosis, just like a candle wax ("Candle Wax Sign" - Figs. 10 and 12). Generally only one side of the bone is compromised. Usually just the x-ray is enough for the diagnosis (Fig. 11), but CT reveals more details of the disease such as cortical and medullar portions of the bone involvement. (Figs. 13-14) Radionuclide bone scanning reveals moderate and asymmetric increased uptake (Fig. 15).

**Tuberous Sclerosis (TS).** The most common radiologic skeletal findings in TS include hyperostosis of the calvaria, osteoblastic or osteosclerotic changes in the spine and cystic changes circumscribed by a sclerotic ring; these last ones are more frequent in the phalanges. In the chest wall we usually see a predominance of these sclerotic bone lesions (SBL) all over the posterior elements of the spine, conforming to the shape of the lamina, pedicles, transverse process, or spinous process (Figs. 16-17). None of these SBL expands the bone, deforms or extends beyond the cortex. Four or more of these SBL are more common in patients with TS and LAM, than in patients with only TS or only LAM. The differential diagnosis of SBL includes osteoblastic metastasis, osteopoikilosis, and mastocytosis. When these sclerotic findings are associated with lung involvement
signs of LAM and MMPH such as lung cysts and micronodules with random distribution (Figs. 18-19), respectively, the hypothesis of TS should be raised.

**Hereditary multiple exostoses (HME).** This syndrome is characterized by a vast quantity of osteochondromas. They generally grow in the knees, ankles and shoulders. In the chest they are rare and when present they can affect the ribs, clavicles and scapulae (Figs. 20-21). The most important characteristic feature of osteochondromas is uninterrupted merging of the cortex of the host bone with the cortex of the lesion and the communication of the medullary portion of the exostoses and the medullary cavity of the host bone. The CT scanning can show unequivocally all these details.
Fig. 0: Middle-aged woman with SAPHO Syndrome. Axial MDCT image shows sternoclavicular joints with osteosclerosis and ankylosis of the joint space.

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Fig. 0: Middle-aged woman with SAPHO Syndrome. Volume Rendering image shows sternoclavicular joints with osteosclerosis and ankylosis of the joint space.

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Fig. 0: Middle-aged woman with SAPHO Syndrome. Volume Rendering image shows sternoclavicular joints with osteosclerosis and ankylosis of the joint space.

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Fig. 0: Young female with Poland's Syndrome. CT Scout shows unilateral hyperlucency with absence of the normal axillary fold on the right that mimics a radical mastectomy. Hypoplasia of the ipsilateral third and fourth ribs is also noticed.
Fig. 0: Young female with Poland's Syndrome. CT Volume Rendering image shows aplasia of nipple and breast and absence of the greater and smaller pectoral muscles. Hypoplasia of the ipsilateral third and fourth ribs is also noticed.

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Fig. 0: Young female with Poland’s Syndrome. CT Volume Rendering image shows hypoplasia of the ipsilateral third and fourth ribs.

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Fig. 0: Young female with Poland's Syndrome. Axial MDCT image shows absence of the serratus anterior on the right (green arrow) and presence of this muscle on the left side (red arrow).

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Fig. 0: Young female with Poland's Syndrome. Axial MDCT image shows absence of the greater and smaller pectoral muscles on the right (green Arrow) and presence of these ones on the left side (red arrow).

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**Fig. 0:** Young female with Poland's Syndrome. Axial Volume Rendering image shows absence of the greater and smaller pectoral muscles on the right and presence of these ones on the left side.

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**Fig. 0:** Middle-aged woman with Melorheostosis. A rib compromised showing the "candle wax sign" at the superior border of the rib. Only one side is compromised, the superior one.

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**Fig. 0:** Middle-aged woman with Melorheostosis. Digital X-Ray showing a compromised rib at the right rib cage.

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**Fig. 0:** Middle-aged woman with Melorheostosis. CT Volume Rendering showing a rib compromised at the right rib cage.

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Fig. 0: Middle-aged woman with Melorheostosis. Axial MDCT image showing a rib compromised at the right rib cage (green arrow). Compared with a normal rib, at the left side (red arrow), look at the cortical and medullar portions of the bone involvement.

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**Fig. 0:** Middle-aged woman with Melorheostosis. Coronal MDCT image showing a rib compromised at the right rib cage (green arrow). Compared with a normal rib, at the left side (red arrow), look at the cortical and medullar portions of the bone involvement.

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Fig. 0: Middle-aged woman with Melorheostosis. Radionuclide bone scanning reveals asymmetric increased uptake.

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Fig. 0: Young adult female with tuberous sclerosis. Sagital MDCT image showing some sclerotic bone lesions all over the posterior elements of the spine, conforming to the shape of the pedicles and articular processes (green arrow).

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Fig. 0: Young adult female with tuberous sclerosis. Axial MDCT image showing a sclerotic bone lesion conforming to the shape of the pedicle of the thoracic spine (green arrow).

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Fig. 0: Young adult female with tuberous sclerosis. Axial MDCT image showing micronodules with random distribution (green arrow), compatible with multifocal micronodular pneumocyte hyperplasia (MMPH).

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**Fig. 0:** Young adult female with tuberous sclerosis. Coronal MDCT image showing micronodules with random distribution (green arrow), compatible with multifocal micronodular pneumocyte hyperplasia (MMPH).

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Fig. 0: Young female with hereditary multiple exostoses. Volume Rendering CT image shows a vast quantity of osteochondromas distributed in the ribs, clavicles and scapulae (green arrows).

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Fig. 0: Young female with hereditary multiple exostoses. Axial Volume Rendering CT image shows a osteochondroma of the rib with intrapulmonary growth through a accessory fissure (green arrow).

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Conclusion

A vast variety of syndromes have associated chest wall bones alterations. Some of them are very peculiar. These changes can be associated with other findings in lungs, soft tissue and the partial section of the superior abdomen, included in the chest study. With all those findings together the radiologist may be able to suggest a syndromic diagnosis. Therefore, every radiologist should take a good and careful look at the chest wall, and don't forget that the chest is much more than just heart and lungs.
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References