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Clinical Image

Calcified Atherosclerosis of the Pulmonary Trunk, Stenosis of the Main Pulmonary Arteries, and Post-Stenotic Dilation of Segmental Pulmonary Arteries in a Patient With Alagille Syndrome☆

Aterosclerosis calcificada del tronco de la arteria pulmonar, estenosis de las arterias pulmonares principales y dilatación postestenótica de arterias pulmonares segmentarias en un paciente con síndrome de Alagille

Luis Gorospe Sarasúa,* Ana María Ayala-Carbonero, María Ángeles Fernández-Méndez

Servicio de Radiodiagnóstico, Hospital Universitario Ramón y Cajal, Madrid, Spain

We report the case of a 48-year-old man with a history of Alagille syndrome (AS) who consulted due to dyspnea and chest discomfort. The patient had undergone a liver and kidney transplant in the past. Chest radiography revealed an increase in the caliber of several pulmonary arteries in the right lung base (Fig. 1A). A chest computed tomography showed calcified atheromatous plaques in the pulmonary artery trunk (Fig. 1B), stenosis of the main pulmonary arteries (Fig. 1C), and post-stenotic dilation of the segmental pulmonary arteries of the right lower lobe (Fig. 1D).

AS is a rare autosomal dominant genetic disease that particularly affects the liver, the kidneys and the heart. In the lungs, AS is characteristically associated with pulmonary artery stenosis, which can cause pulmonary hypertension and right heart failure in the long term.[1] Currently, this stenosis can be treated with open surgery or angioplasty with stent placement. We found scant reference in the literature to post-stenotic dilation of the peripheral pulmonary arteries in AS.[2]

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* Corresponding author.
E-mail address: luisgorospe@yahoo.com (L.G. Sarasúa).

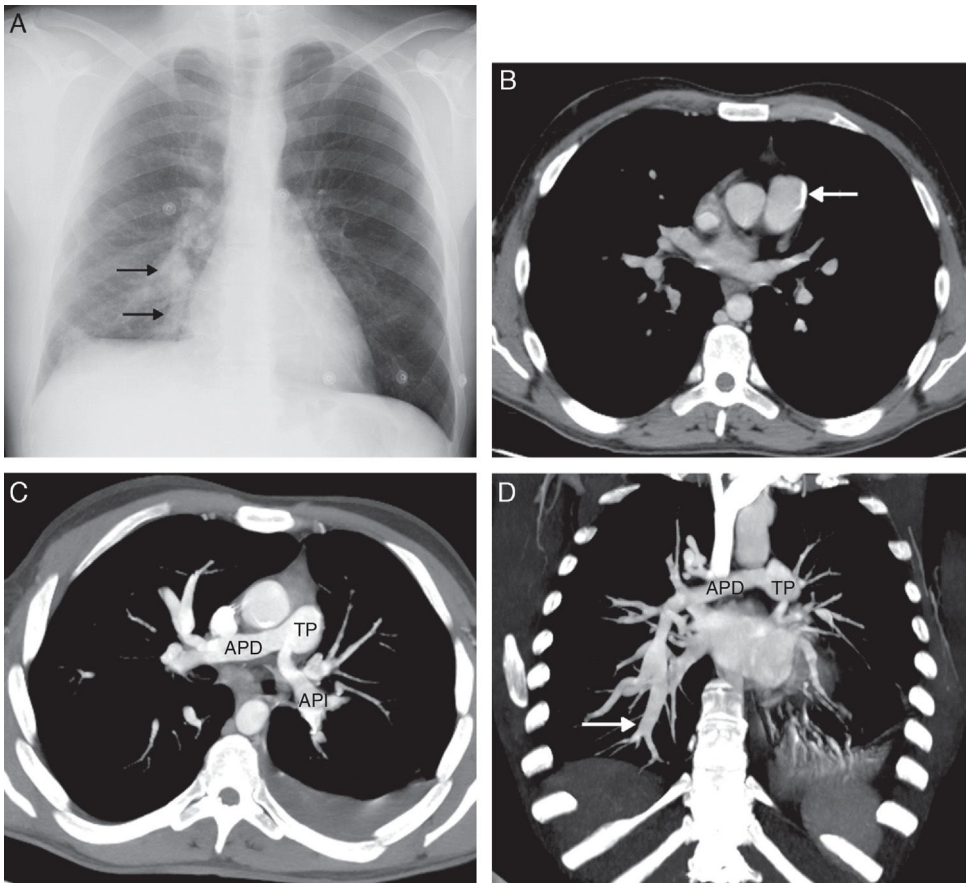

Fig. 1. (A) Posteroanterior chest radiograph, showing increased caliber of arteries in the right lung base (arrows). Note the oligemia in the left lung compared to the right. (B) Axial image of chest CT, showing a calcified atheromatous plaque (arrow) in the wall of the pulmonary artery trunk. (C) Axial maximum intensity projection (MIP) reconstruction of chest CT, showing stenosis of the main pulmonary arteries, particularly in the left side (right main pulmonary artery [APD]; left main pulmonary artery [API]; pulmonary artery trunk [TP]). (D) Coronal MIP reconstruction of chest CT showing segmental artery in right lower lobe (arrow) of larger caliber than the APD (APD, TP).

References

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