



Clinical Image of the Month

Stanford B or DeBakey IIIa complicated aortic dissection in a patient with Ehlers-Dals syndrome type IV

Diseción aórtica complicada de tipo Stanford B (o DeBakey IIIa) en paciente con síndrome de Ehlers-Danlos de tipo IV

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CASE REPORT

A 34-year-old hypertensive male, poorly controlled, with a history of Ehlers-Danlos syndrome type IV and no cardiovascular signs presented with sudden chest and interscapular pain. Physical examination revealed blood pressure levels of 184/96 mmHg, a hollowed chest, and brachydactyly in the patient's left hand. Chest x-ray showed mediastinal widening, and the

panoramic oral x-ray performed revealed the presence of supernumerary teeth (Fig. 1). Thoracic coronary computed tomography angiography revealed the presence of an intimal flap and distal dissection trace to the left subclavian artery, classified as Stanford type B (DeBakey type IIIA) (Fig. 2). Given the classification and hereditary association, the heart team decided to go implant a stent-graft. At the 6-month follow-up, the patient remains asymptomatic with good graft patency.

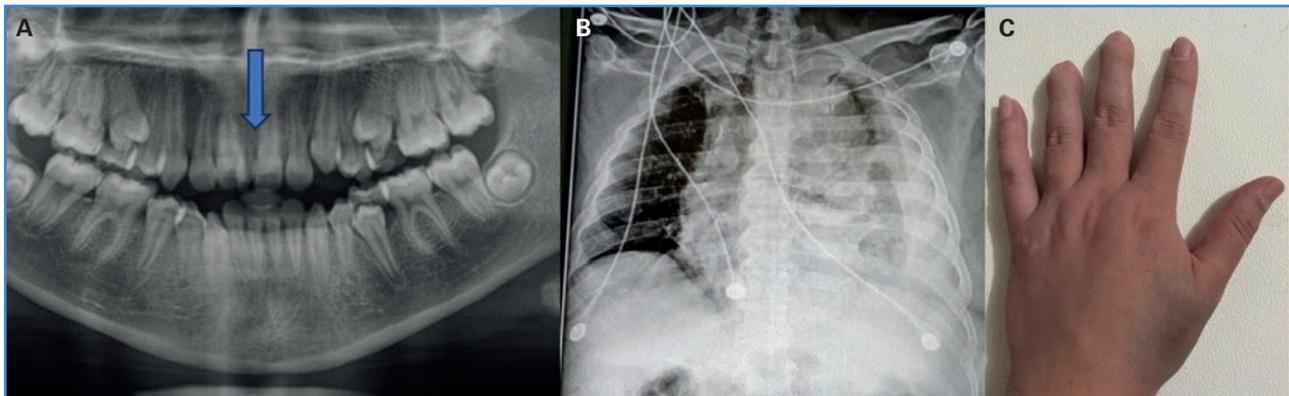


Figure 1. A. Panoramic radiograph showing interincisal supernumerary tooth. B. Chest radiograph. Mediastinal widening and grade III cardiomegaly. C. Brachydactyly in 3rd and 4th toes with diffuse ecchymosis.

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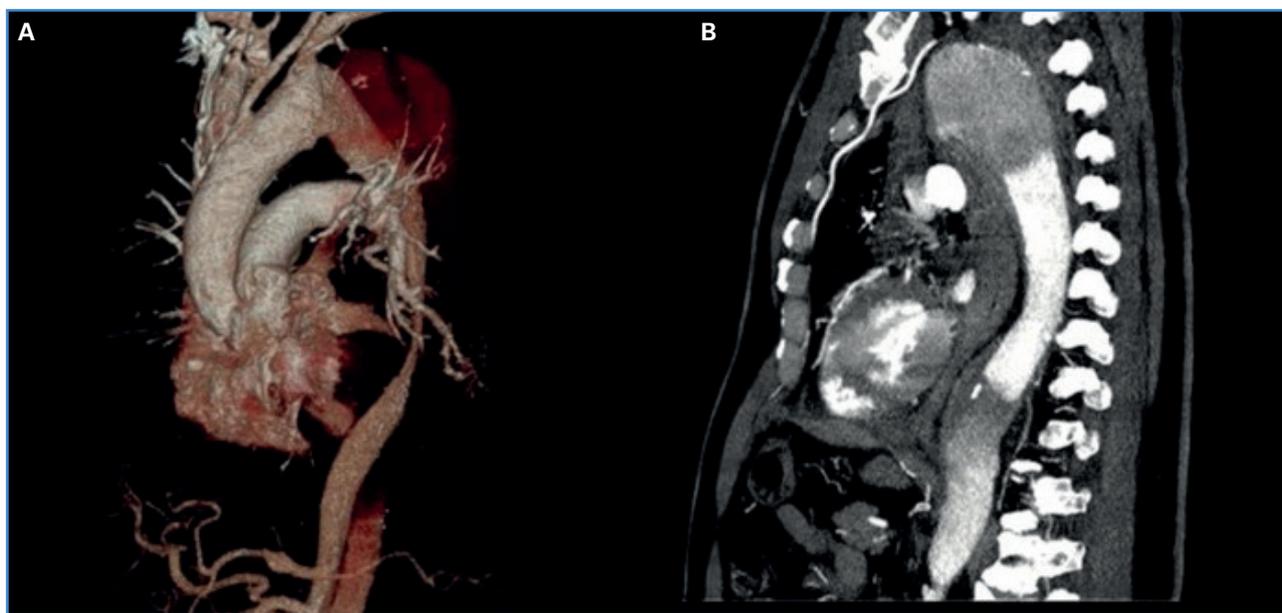


Figure 2. A. 3D angiotomography. Dilatation at the emergence of the descending thoracic aorta with presence of dissection trace distal to the subclavian artery. B. Contrast-enhanced chest CT scan showing aortic dilation distal to the left subclavian.

DISCUSSION

Although aortic dissection is rarely associated, when it does it comes with high morbidity and mortality rates, along with connective tissue disorders characterized by collagen changes and tissue metabolism, such as Ehlers-Danlos syndrome type IV, which has a characteristic phenotype: brachydactyly, supernumerary teeth, and spontaneous ruptures with minimal trauma; mortality occurs due to vascular rupture (1-3).

Back in 2014, the European Society of Cardiology (ESC) recommended thoracic endovascular aortic repair (TEVAR) for type B aortic dissection complicated by syndromic factors, as in this case (class IC) (4).

The INSTEAD trial documented the effectiveness of TEVAR in patients with type B aortic dissection associated with connective tissue disorders, which improves aortic remodeling more than medical treatment alone, or surgery (5).

According to our report, aortic dissection with hereditary association and connective tissue disorders,

and a high risk of complications presents a suitable scenario for stent-graft implantation, a safe procedure that improves the patient's prognosis.

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