Right-sided chest pain in Poland syndrome

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Poland syndrome is a rare congenital abnormality with an estimated incidence of 1 in 20,000 to 30,000 live births.\(^1\) It involves unilateral hypoplasia of the major pectoralis muscle and brachysyndactyly of the ipsilateral upper extremity. The pathogenesis involves reduced blood flow to the subclavian and vertebral arteries during early fetal development.\(^2\) A 3-to-1 male predominance is typically seen\(^3\) and three-quarters of cases involve the right hemithorax and upper extremity.\(^4\)

The association of dextrocardia with left-sided Poland syndrome is very rare, with only 20 cases thus far described in the literature. A handful of those cases have involved rib defects and pain on the left side.\(^1,4–7\) Until now, there have been no reports of right-sided chest pain in the setting of left-sided Poland syndrome.

Case report

A 46-year-old Australian man with a “right-sided heart” presented to an outpatient clinic with a 5-year history of dull, constant right-sided chest pressure. He denied shortness of breath, palpitations, nausea, diaphoresis, or cough. He reported that the pain improved with aspirin and worsened with rotation of his torso.

The patient recalled that he was overdue at birth and associated his traumatic delivery with his anatomic anomalies. He was convinced that surgery had been performed on his left hand, which resulted in his short fingers.

He was told his heart was located on the right side of his chest. He was taking no medications, was unemployed, and suffering from anxiety. Both of his sisters had diabetes mellitus and many family members were obese; no family history of heart disease, dextrocardia or congenital disease.

On physical examination, his pulse was 100 beats per minute and blood pressure was 149/93 mmHg. He weighed 119.3 kg (body mass index 29) with an abdominal circumference of 102 cm. He had a hypoplastic left chest, with an absent nipple and pectoral muscle (Figure 1), brachydactyly of the left hand (Figure 2), and a hypoplastic left upper extremity.

His haemoglobin A1c was 10.9%, fasting total cholesterol 22.5 mg/L, triglycerides 28 mg/L, high-density lipoprotein 3.4 mg/L, and low-density lipoprotein 14.3 mg/L. A right-sided electrocardiogram revealed sinus tachycardia. A chest X-ray showed dextrocardia with a left-sided aortic knob and a left-sided gastric bubble (Figure 3). A stress echocardiogram showed normal right and left-sided chamber size and function, but the acquisition of the images was to the right of the midline.
Figure 1. Hypoplastic left chest, absent left nipple and absent left pectoral muscle

Figure 2. Brachydactyly of the left hand
The patient was informed of the diagnosis of Poland syndrome and was reassured that his chest pain was most likely musculoskeletal in origin. He was also informed that he had metabolic syndrome, which placed him at higher risk for cardiovascular disease. He subsequently improved on anti-inflammatory therapy, but was instructed to seek emergency care should his chest pain recur.

**Discussion**

This case illustrates the potential to miss a diagnosis of cardiac ischaemia in a patient with an anatomic anomaly who presents with atypical chest pain. Although less likely ischaemic chest pain, clinical suspicion of acute coronary syndrome in this patient was maintained as he had concurrent metabolic syndrome. Pooled data from 37 studies of more than 170,000 patients have shown that metabolic syndrome doubles the risk of coronary artery disease.\(^8\)

While there have been reports of left-sided chest pain in patients with left-sided Poland syndrome and associated dextrocardia,\(^1,4-7\) this is the first known case of right-sided musculoskeletal pain in this setting.

The patient in this case has dextroposition of the heart but an otherwise normal stress echocardiogram. In the setting of metabolic syndrome, however, ischemia should be considered in the differential diagnosis for right-sided chest pain with left-sided Poland syndrome, given the increased risk for cardiovascular disease.
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References: