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(CASE REPORT)

Teratoma of the pericardium revealed by seronegative polyarthritis: A case report

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Abstract

Introduction: Rheumatoid arthritis (RA) is an inflammatory, autoimmune disease with preferential involvement of the synovium in the proximal joints of the hands and feet. Formerly known as chronic progressive polyarthritis or chronic rheumatoid arthritis, its course is characterised by joint erosion and destruction. Coronary heart disease and strokes, the consequences of atheromatous disease, are part of rheumatoid arthritis and are responsible for the increased morbidity and mortality observed in this disease.

Cardiac tumours in children are rare. The frequent use of echocardiography has helped to detect them earlier and with greater sensitivity. Most cardiac tumours in children are congenital, primary and benign. Most are discovered in children under one year of age.

The aim of this study is to report the discovery of this type of tumour in a 10-year-old child being treated for seronegative polyarthritis.

Observation: We report the observation of a 10-year-old girl, from a non-consanguineous marriage, with a history of systemic arthritis on difal 25mg *2/d + MTX 0.8mg/kg/week. Present since the age of 3 with recurrent arthralgias, admitted for polyarthralgias, fever with chest pain and whose examination found swelling, pain and limitation of passive and active movement of the left knee joint and elbow joint. Biological work-up revealed inflammatory microcytic hypochromic anaemia, normal ferritin, elevated CRP and ESR, ECBU, stool culture, ASLO serology, sputum BK, sputum TB PCR, Gene Xpert in sputum, Quantiiferon, SAM work-up and rheumatoid factors (RF) were negative, antinuclear antibodies (ANA) positive.

EX. Ophthalmology and abdominal ultrasound normal, chest X-ray reveals cardiomegaly (ICT 0.6), X-ray of the spine + knee + elbow showed a discreet increase in the transparency of the bone structure. ETT found an oval mass measuring 20/25mm at the level of the junction between OD and VD not managing the reflux of the tricuspid valve and THORACIC CT in favour of a mass adherent to the pericardium evoking in the first instance a teratoma.

Conclusion: Cardiac tumours in children are rare, and pericardial teratomas are often benign. In this study, we emphasise the importance of looking for underlying cardiac disease in the presence of recurrent episodes of arthralgia in children. Early positive diagnosis of the disease and the therapeutic implications are vital.

Keywords: Rheumatoid arthritis; Polyarthritis; Oval mass; Pericardial teratoma

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1. Introduction

Rheumatoid arthritis (RA) is an inflammatory autoimmune disease with preferential involvement of the synovium in the proximal joints of the hands and feet. Formerly known as chronic progressive polyarthritis or chronic rheumatoid arthritis, the disease is characterised by joint erosion and destruction (1).

Critical appraisal of the clinical use of serological tests in RA has highlighted the importance of seeking diagnoses other than RA in seronegative patients (2).

Functional disability in RA is the result of chronic inflammation of the joint synovial membrane, leading to progressive destruction of bone and cartilage. Extra-articular manifestations of the disease can be life-threatening. Coronary heart disease and stroke, the consequences of atheromatous disease, are part of rheumatoid arthritis and are responsible for the increased morbidity and mortality observed in this disease.

Cardiac tumours in children are rare. The frequent use of echocardiography has helped to detect them earlier and with greater sensitivity. Most cardiac tumours in children are congenital, primary and benign. Most are discovered in children under one year of age.

Teratomas are tumours containing differentiated tissue from 1, 2 or all 3 primordial germ layers (endoderm, mesoderm and ectoderm) developed from the omnipotent cells of Hensen's node, near the allantois and the yolk bladder. These primordial germ cells migrate from the 4th week along the dorsal mesentery towards the genital crest where they arrive around the 6th week and transform into gonads. Stopped migration or aberrant migration is thought to be the cause of the development of extra-gonadal tumours.

Teratomas may develop in the anterior mediastinum (rarely posterior) and in the pericardial space. The appearance is variable and once again resembles lymphangiomas. Their prognosis is relatively good (3, 4).

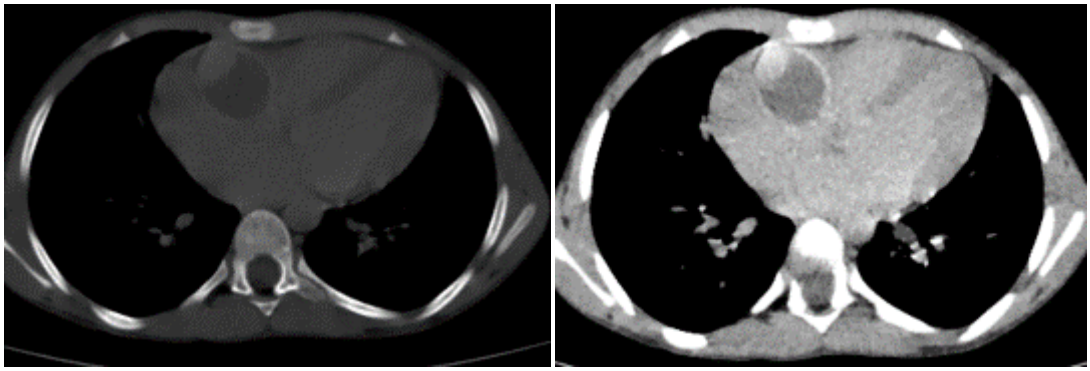
Standard X-rays combined with chest CT scans are sufficient to diagnose these tumours.

Surgical removal of these tumours as completely as possible remains the only conceivable therapeutic approach in order to avoid complications and local recurrence (5, 6, 7).

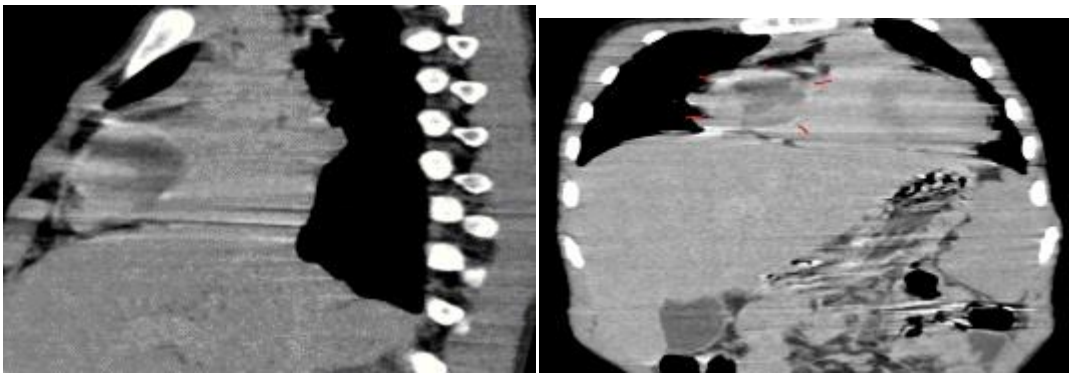
2. Observation

We report the observation of a 10-year-old girl, from a non-consanguineous marriage, with a history of systemic arthritis on difal 25mg *2/d + MTX 0.8mg/kg/week. She had been suffering from recurrent arthralgia since the age of 3 and was admitted with polyarthralgia, fever and chest pain. On examination, she was found to have swelling, pain and limitation of passive and active movement in the left knee joint and elbow joint. Biological work-up showed inflammatory microcytic hypochromic anaemia, normal ferritin, elevated CRP and ESR, ECBU, stool culture, ASLO serology, sputum BK, sputum TB PCR, Gene Xpert in sputum, Quantiiferon, SAM work-up and rheumatoid factors (RF) were negative, antinuclear antibodies (ANA) positive.

EX. Ophthalmology and abdominal ultrasound normal, chest X-ray revealed cardiomegaly (ICT 0.6), X-ray of the spine + knee + elbow showed a discreet increase in the transparency of the bone structure, respecting the joint spaces and no suspicious osteolytic or osteocondensing lesion visible and respecting the molar parts. ETT found an oval mass measuring 20/25mm at the level of the junction between the OD and VD not managing the reflux of the tricuspid valve and thoracic CT in favour of a mass adherent to the pericardium, suggesting a teratoma in the first instance (Figure 1).



a) Axial section



b) sagittal section C) coronal section

Figures 1 Chest scan in favor of a Teratoma (a, b, c)

3. Discussion

Rheumatoid arthritis (RA) is an inflammatory, autoimmune disease with preferential involvement of the synovium in the joints of the limbs, causing cartilage and bone destruction and ultimately functional disability (8).

Germ cell tumours are rare tumours. They are located in the gonads in over 80% of cases. The most common extragonadal location is mediastinal, giving rise to germinal tumours of the mediastinum (9).

In practice, germ cell tumours of the mediastinum are classified into three groups: seminomas, non-seminomatoustumours and teratomas.

A teratoma is a germ cell tumour derived from one or more of the three primitive embryonic layers: endoblast, mesoblast and ectoblast. It is made up of various tissues whose appearance is reminiscent of the different stages of embryonic development, up to the adult stage in some cases, tissues which are normally foreign to the organ or anatomical region in which they are found.

Mature teratomas are the frequent benign variety of germ cell tumours of the mediastinum (70% in children and 60% in adults) (9). These tumours are primarily manifested by symptoms associated with their impact on the various anatomical structures of the mediastinum. Computed tomography is the gold standard for diagnosing these tumours (10).

The primitive germ cells can proliferate into benign lesions (mature teratomas and grade 1 immature teratomas), malignant lesions (high grade immature teratomas and malignant germ cell tumours) or benign lesions and, exceptionally, secondary malignant lesions (cancerised teratomas).

Teratomas can be discovered at any age, from foetus to child, but also in adulthood. In the foetus, many cases are detected as a result of the systematic use of obstetric ultrasound scans. In children, tumours are diagnosed following complications linked to the size of the tumour.

Mediastinal teratomas are diagnosed following chest pain or cardiorespiratory distress (compression, pleural or pericardial effusion). They will appear on imaging as large masses in the anterior mediastinum, much more rarely in the posterior mediastinum, containing tissue of varying density and calcifications (11, 12).

Benign mature teratomas are the most common germ cell tumours in the thorax. Complete surgical removal of these tumours can be very difficult because of the relationships and adhesions with the large vessels and the pericardium, and possible complications such as rupture (12).

Standard X-rays combined with chest CT scans are sufficient to diagnose these tumours.

The only treatment for mature teratomas of the pericardium is complete surgical removal (5, 6, 13, 7, 14, 15, 16, 17).

This treatment has a triple benefit: removal of the mass, confirmation of the diagnosis and prevention of complications. It should never be postponed because of possible local complications and, above all, because of the possible development of a malignant contingent (18, 19).

Detailed anatomopathological examination of the surgical specimen is the only way of making a definitive diagnosis and ruling out the presence of an immature contingent or a frankly malignant focus with an often dreadful prognosis.

The prognosis of teratomas will depend on their composition (mature or immature teratoma, pure or associated with a malignant germ cell component), location, tumour impact, possible complications (rupture, adhesions, haemolyticanaemia, etc.) and the possibility of complete surgical removal (4, 20).

4. Conclusion

Cardiac tumours in children are rare, and pericardial teratomas are often benign. In this study, we emphasise the importance of looking for underlying cardiac disease in the presence of recurrent episodes of arthralgia in children. Early positive diagnosis of the disease and the therapeutic challenge (surgical removal) are essential.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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