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Clinical Case Report

A rare case of regressively changed lipomatous hypertrophy of the interatrial septum presenting with anemia and recurrent fever

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ABSTRACT

Lipomatous hypertrophy of the interatrial septum (LHIS) is characterized by excessive accumulation of adipose tissue within some segments of the interatrial septum. Only one published case so far describes fever as a presenting feature of LHIS. On the other hand, systemic symptoms including anemia and fever are well-known clinical presentations of cardiac myxomas. We report an unusual case of a 79-year-old woman who was thoroughly but unsuccessfully investigated for recurrent fever and anemia in several specialized departments over the course of 4 years. Computed tomography scan showed a pathological mass localized in the interatrial septum and spreading to ascending aorta. Histological analysis of the biopsy samples from surgery revealed the unexpected diagnosis of regressively changed LHIS. We discuss the clinical and pathologic features of this lesion suggesting that its regressive changes may be associated with inflammation and can cause systemic symptoms such as fever and anemia.

1. Introduction

Lipomatous hypertrophy of the interatrial septum (LHIS) is characterized by excessive accumulation of adipose tissue within some segments of the interatrial septum (IAS). The etiology as well as the clinical classification of this lesion remains unclear [1]. LHIS is a rare but increasingly recognized lesion. Its prevalence is estimated to be around 2.2% in the general population [2]. Although LHIS is often an incidental finding in asymptomatic patients, this lesion may be associated with clinical complications, especially supraventricular arrhythmias [3]. Although systemic symptoms including anemia and fever are well-known clinical manifestations of cardiac myxomas, only one published case describes fever as a presenting feature of LHIS [4].

2. Case description

A 79-year-old female was thoroughly but unsuccessfully investigated for recurrent fever and mild to moderate anemia of unknown etiology in several specialized departments between the years 2010 and 2014. Her past medical history was negative for cardiac symptoms including syncope. Besides routine examination methods, this investigation included gastroscopy, computed tomography (CT) colonoscopy, magnetic resonance of the abdomen and pelvis, and fluorescence activated cell sorting of peripheral blood and bone marrow biopsy, as well as repeated laboratory examinations including hemocultures, analysis of autoantibodies, and tumor markers. In September 2014, the patient was hospitalized again for progression of anemia, and at that time, CT examination of the chest and abdomen revealed a pathological mass infiltrating the IAS of the heart. Subsequently, she was referred to our department for further specialized cardiologic examination. Except for pale skin and body temperature in the subfebrile range (37.4°C), the physical examination was unremarkable. Electrocardiogram showed sinus rhythm, with broad biphasic P waves seen in the inferior leads as the only abnormality. Laboratory examination revealed microcytic hypochromic anemia with a hemoglobin level of 87 g/L but a normal level of ferritin 150 μg/L, suggesting anemia of chronic disease. Repeated examinations showed persistent elevation of C-reactive protein ranging from 74 to 107 mg/L and erythrocyte sedimentation rate (Fahraeus-Westergreen test: 120 mm/h), in contrast to a normal level of procalcitonin (0.04 ng/ml). Moreover, blood cultures were repeatedly negative. Transthoracic echocardiography (TTE) showed a well-demarcated, voluminous pathologic mass infiltrating the whole IAS measuring 41 mm×53 mm×56 mm. The lesion spread from the IAS toward the aortic root without signs of obstruction or invasion to the aorta. The mass had a hyperechogenic marginal zone and hypoechogenic

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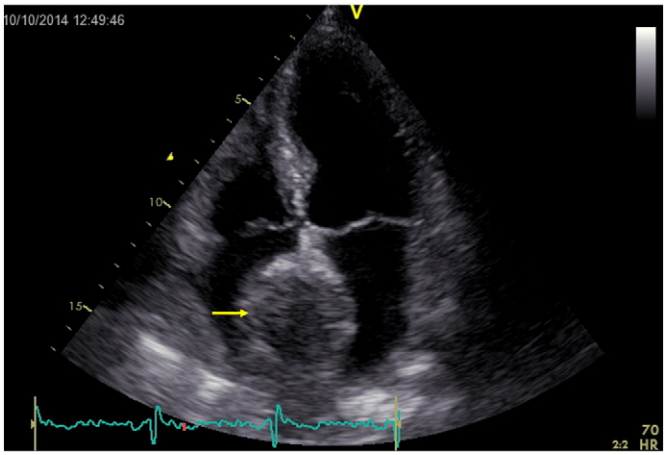

Fig. 1. TTE showed a well-demarcated, voluminous pathologic mass measuring 41 mm×53 mm×56 mm and infiltrating the whole IAS (yellow arrow) in the apical four-chamber view. The mass exhibits a hyperechogenic margin and hypoechogenic central parts in contrast to the myocardium. The typical "dumbbell"-shaped configuration characteristic for LHIS is absent.

central parts in contrast to the myocardium. The typical "dumbbell"-shaped configuration characteristic for LHIS was absent (Fig. 1). Cardiac magnetic resonance (CMR) confirmed the presence of a well-demarcated oval-shaped lesion in the IAS, which was hypointense on BTFE-CINE imaging. The lesion had heterogeneous high-signal intensity on fat-suppressed T2-weighted images and low-signal intensity on T1-weighted black blood images, with heterogeneous enhancement after administration of gadolinium contrast agent (Fig. 2A–D). CT showed a polycyclic soft-tissue mass measuring 50 mm×53 mm×60 mm. Similar to TTE findings, the mass spread from the IAS to the ascending aorta, which was not infiltrated or compressed. The center of the mass was hypodense with an average density of 44 HU (Hounsfield units) and did not enhance after ionic contrast administration. In comparison with the central zone, peripheral parts of the lesion had average density of 45 HU and enhanced after intravenous contrast administration (108 HU). Positron emission tomography–computed tomography (PET-CT) described this mass as a hypermetabolic tumor expansion infiltrating the IAS with increased fluorodeoxyglucose (FDG) uptake in the peripheral zone of the lesion (Fig. 3). The CT and PET-CT appearance of the mass suggested regressive changes in its center. Since these imaging findings raised suspicion for a malignant tumor located in the IAS, we performed percutaneous transvenous biopsy, the results of which were unfortunately inconclusive. Therefore, cardiac surgery was indicated. However, the mass proved to be inoperable due to its size and relation to the aortic root, and only biopsy samples were obtained. Histological examination of the two biopsy samples from the mass revealed unencapsulated mature adipose tissue permeated with pronounced fibrous trabeculae as well as partially hyalinized dense collagenous tissue (Fig. 4A and B). There was intense inflammatory infiltration (CD3+ and CD20+ lymphocytes; kappa+ and lambda+ plasma cells) localized mainly in the surface layer of the biopsy sample (Fig. 4C). Furthermore, the presence of hemosiderophages indicated bleeding. Cardiomyocytes were entrapped sporadically

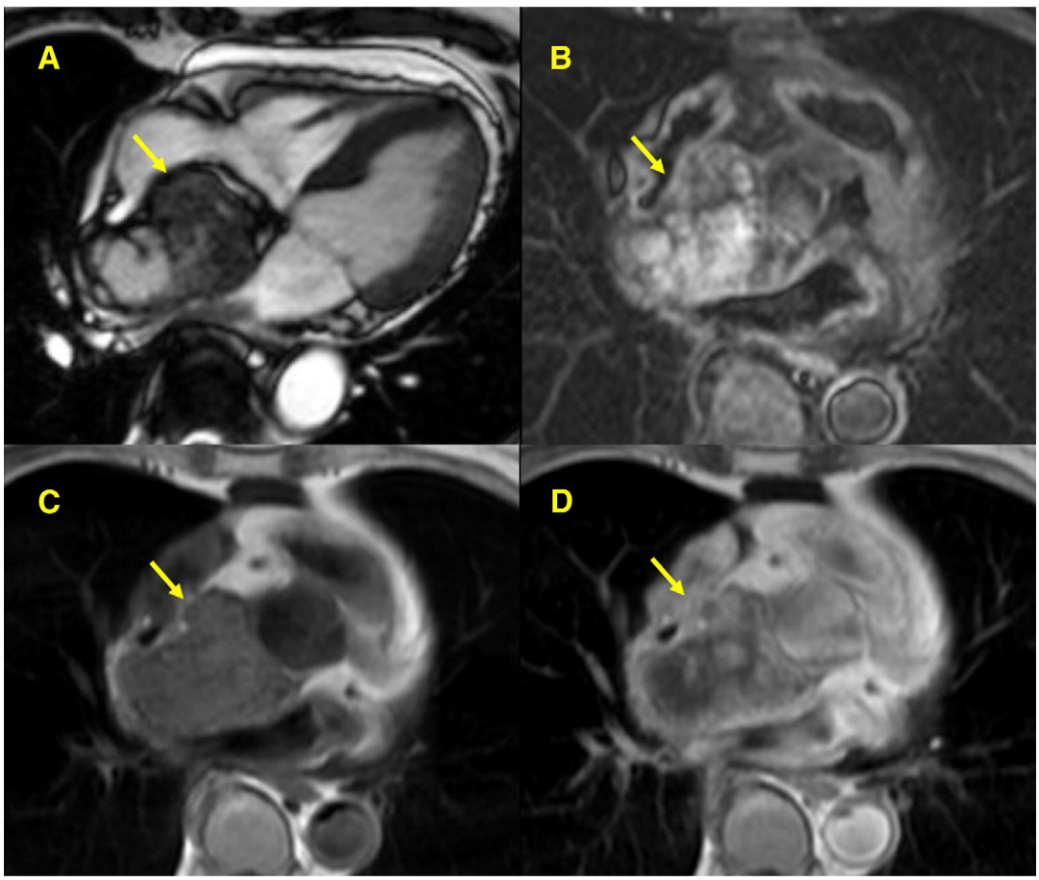

Fig. 2. (A–D) CMR images displayed a well-demarcated oval lesion measuring 50 mm×52 mm×66 mm localized in the IAS (yellow arrows). The mass was hypointense on BTFE-CINE sequences (A) had heterogeneous high-signal intensity on fat-suppressed T2-weighted images (B) and low-signal intensity on T1-weighted black blood images (C) with heterogeneous enhancement after administration of gadolinium contrast agent (D).