

Case report: multi-modality imaging of a right ventricular fibroma in a teenager

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Background

Cardiac fibroma is a rare primary benign tumour of the heart. It often causes arrhythmia, endangers the lives of patients, and has a worse prognosis than other benign tumours. We report a 14-year-old female patient with a right ventricular fibroma. Various preoperative imaging examinations showed that the lesion was benign, and postoperative pathology confirmed that the lesions were fibroma.

Case summary

A 14-year-old female patient visited her doctor for more than 5 months because of a heart murmur. Echocardiography revealed a slightly hyperechoic mass in the right ventricle, and on myocardial perfusion contrast imaging, the lesion showed equal enhancement. And the lesion also showed enhancement on contrast-enhanced gated cardiac computed tomography (CT). Contrast-enhanced magnetic resonance imaging (MRI) of the heart revealed that the lesion was isointense on T₁-weighted image (T₁WI), and isointense to slightly hyperintense on T₂-weighted image (T₂WI). The lesion was significantly homogeneously enhanced on a delayed enhancement scan. A positron emission tomography-CT (PET-CT) with 18F-fluorodeoxyglucose (18F-FDG) demonstrated that the mass showed lower levels of 18F-FDG uptake. These features suggested this lesion was a benign lesion. The postoperative pathology suggested the lesion was a right ventricular fibroma. The patient was discharged 14 days after surgery and remains disease-free and asymptomatic 14 months after surgery.

Discussion

Cardiac fibromas are histologically benign, but they can cause obstruction and malignant arrhythmia. The gold standard for diagnosing fibroma is pathology. However, in the absence of pathology, it is necessary to use various imaging methods to evaluate the lesions to distinguish between benign and malignant tumours.

Keywords

Primary cardiac fibroma • Multi-modal imaging • Arrhythmia • Surgical resection • Case report

ESC Curriculum

2.2 Echocardiography • 7.5 Cardiac surgery • 2.2 Echocardiography • 7.5 Cardiac surgery

Learning points

- Cardiac fibroma is a pathologically benign tumour, which tends to cause malignant arrhythmia and has a poorer prognosis than other benign tumours.
- Preoperative evaluation of lesions by multi-modal imaging and early resection of lesions are the key to improve the prognosis of patients.

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Introduction

Cardiac tumours are relatively rare with a total incidence of <0.33%. Primary cardiac fibromas account for 1% of adult primary cardiac tumours.¹ Cardiac fibromas are histologically benign, but they can cause obstruction and arrhythmia.² If not handled properly, the mortality rate is high. In the absence of pathology, it is necessary to use multi-modal imaging to assess the lesions, to distinguish between benign and malignant tumours, and to perform early surgical intervention.

We examined the case of a 14-year-old female patient with a right ventricular fibroma. We diagnosed it as a benign lesion by various examination methods before operation and performed a surgical resection. The pathology after the operation confirmed that the lesion was a fibroma. Early resection of fibroma effectively avoids the risk of sudden death due to arrhythmia. This case report suggests that multi-modal imaging can help us identify the nature of heart tumours, and can provide effective information for the evaluation of patients before and after surgery.

Timeline

Day 1	The patient was hospitalized due to heart murmur found in physical examination. Electrocardiogram suggested incomplete right bundle branch block. Transthoracic echocardiography showed slight hyperechoic mass (about 26 mm × 17 mm) in the right inner wall, nature to be investigated.
Day 76	Gated cardiac computed tomography (CT) enhancement identified an irregular low-density lesion is seen in the right ventricle. Computed tomography value is ~33 HU. Enhanced scan showed enhancement, and the lesion is closely related to the ventricular septum.
Day 88	Positron emission tomography-computed tomography (PET-CT) showed a nodule near the ventricular septum in the right ventricle, no increase in glucose metabolism was observed, and the possibility of benign tumour was considered.
Day 100	Contrast-enhanced magnetic resonance imaging (MRI) showed the lesion showed isointense on T ₁ -weighted image (T ₁ WI) and isointense to slight hyperintense on T ₂ -weighted image (T ₂ WI). The delayed enhanced scan showed obvious homogeneous enhancement. Benign tumour was considered. Fibroma is more likely.
Day 178	The patient was admitted for surgery. Myocardial perfusion angiography showed the

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	perfusion of ultrasonic enhancement agent in the right ventricle mass showed equal enhancement, and the possibility of benign lesions was considered.
Day 180	Complete resection was performed and pathological analysis confirmed the tumour as cardiac fibroma.
Day 194	Patient was discharged.
14 months after discharge	Patient remains disease-free and asymptomatic with no ongoing therapy.

Case presentation

A 14-year-old Asian female patient presented with a heart murmur for more than 5 months. The patient was in good health and had no cardiac family history. A physical examination showed a respiratory rate of 16 breaths/min, heart rate of 70 beats/min (regular), blood pressure of 110/70 mmHg, body temperature of 36.5°C, and percutaneous oxygen saturation of 98% on ambient air. Grade 3 systolic murmur can be heard in the apical region during auscultation. Findings on other cardiovascular system examination were absence.

Electrocardiogram suggests incomplete right bundle branch block. Echocardiography in another hospital revealed that there was an elliptical and slightly hyperechoic mass with a size of 26 × 17 mm in the right inner wall (*Figure 1*), suggesting the possibility of a thrombus. Further examination was recommended. A contrast-enhanced gated cardiac computed tomography (CT) showed that the lesion was closely related to the interventricular septum with enhancement (*Figure 2*). On contrast-enhanced magnetic resonance imaging (MRI), the lesion was isointense on T₁-weighted image (T₁WI) and isointense to slightly hyperintense on T₂-weighted image (T₂WI). The delayed enhanced scan showed obvious homogeneous enhancement (*Figure 3*). Positron emission tomography-CT (PET-CT) examination revealed a nodule in the right ventricle near the ventricular septum with no increase in glucose metabolism, which suggested a high possibility of a benign tumour (*Figure 4*). None of the imaging examinations found any invasion of the surrounding structures by the lesions, no thickening of the pericardium, and no effusion in the pericardial cavity.

To seek surgical treatment, the patient came to our hospital for reexamination by echocardiography and myocardial perfusion angiography. The angiography showed a solid isoechoic mass in the right ventricle, which was closely related to the right ventricle surface in the middle and lower segment of the anterior septum, without an obvious sign of swinging. The perfusion of an ultrasonic enhancement agent in the right ventricle was similar to that in the septal myocardium. After excluding surgical contraindications, a right ventricular mass resection was performed on 2021-04-01. During the operation, the tumour was located in the right ventricular septum. Its pedicle was located near the apex of the ventricular septum (*Figure 5*).

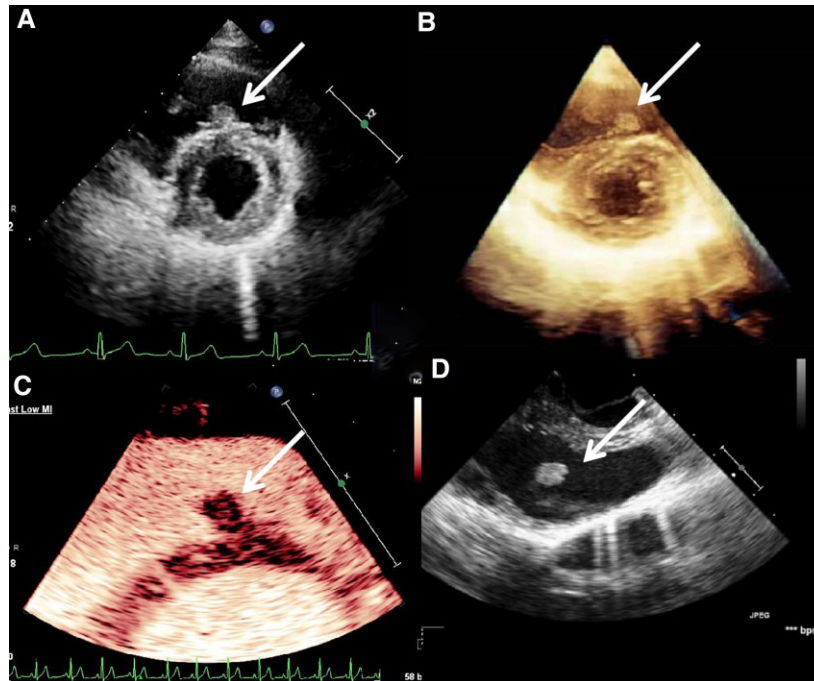


Figure 1 Ultrasound appearance of the lesion. (A) Two-dimensional sonographic image of the lesion, which is located in the right ventricle and appears to be isoechoic (arrow); (B) The spatial location of the lesion is shown by 3D-TTE (arrow); (C) Myocardial contrast echocardiography shows equal enhancement of the lesion (arrow); (D) Transoesophageal echocardiography indicates that the lesion is located in the right ventricle (arrow).

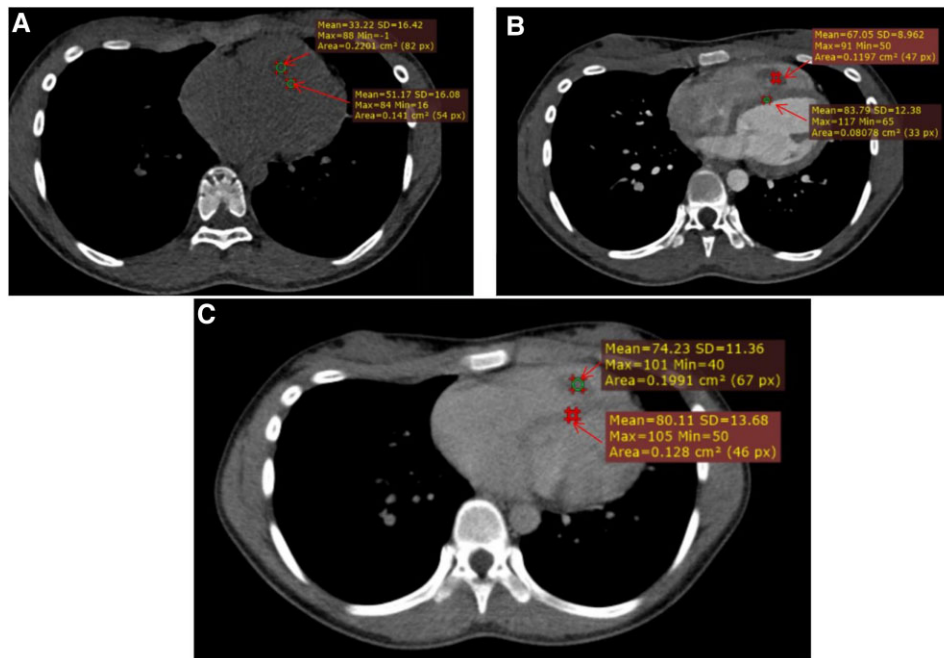


Figure 2 Focus on contrast-enhanced gated cardiac CT: (A) non-contrast scan: an irregular low-density lesion is seen in the right ventricle, CT value is ~33 HU; (B) enhancing scan: the mild enhancement of the lesion, CT value is ~67 HU; (C) delayed scan: the CT value of the lesion was 74 HU.

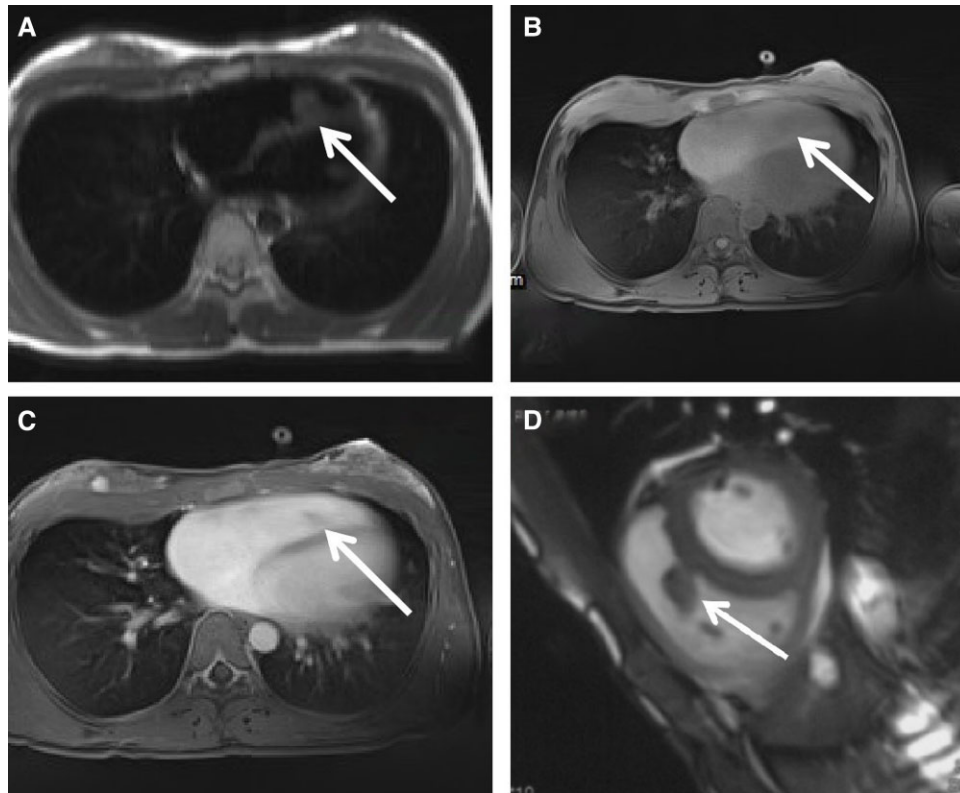


Figure 3 The appearance of the lesion on enhanced MRI. As shown in (A) and (B), the lesion shows isointense on T_1 WI, isointense to slightly hyperintense on T_2 WI (arrow); (C) shows delayed enhancement of the lesion; (D) shows the bright blood SSFP image snapshots (T_2 weighting).

Postoperative pathology confirmed a right ventricle fibroma. Timeline shows a simple procedure for diagnosis and treatment.

The patient was discharged 14 days after surgery and remains disease-free and asymptomatic 14 months after surgery.

Discussion

Cardiac tumours are rare and the total incidence is <0.33%. Primary cardiac fibroma, which accounts for only 1% of primary cardiac tumours in adults, is a benign tumour composed of fibroblasts and connective tissue, mainly occurring in infants and children.^{1,3} Fibroma is more common in the septum and left ventricular free wall, and less common in the right ventricle.

Patients with cardiac fibromas often present with ventricular arrhythmia, cyanosis, dyspnoea, and sudden death. Microscopically, neoplastic cells resemble fibroblasts with variable collagen and elastic tissue, and the margins of the tumour often infiltrate the myocardium.⁴

Fibroma is often seen as a large, non-contractible, and heterogeneous solid mass on echocardiography. Calcification, a relatively common feature on CT, is caused by insufficient blood supply. This feature helps to distinguish fibroma from rhabdomyoma.⁵ There was no obvious calcification on CT in this case, possibly because of the small size of the tumour and adequate blood supply. Myocardial perfusion imaging showed the equal enhancement of

the mass, which may be related to the early growth stage of the tumour. Fibroma usually present with a low signal on T_1 WI and T_2 WI images on magnetic resonance. On a delayed contrast-enhanced scan, the fibroma showed obvious enhancement, which was due to the accumulation of contrast medium in extracellular collagen and elastic fibres.²

The gold standard for diagnosing fibroma is pathology. However, in the absence of pathology, it is necessary to use a variety of imaging methods to assess the lesions and to distinguish between benign and malignant tumours. Echocardiography can show location, size, and number of tumours; the movement of the tumour; the effect of the tumour on hemodynamics and adjacent structures.³ Contrast-enhanced ultrasound can clearly show the outline of a heart mass and dynamically display the blood perfusion process of the disease in real time, which has important reference value for judging the nature of the mass. Unfortunately, the examination scope of this modality is small, and it is not possible to examine multiple areas of the mass at the same time. Because of this limitation, CT and MRI are needed as supplementary scans to provide more comprehensive and accurate diagnostic information. Computed tomography can not only provide anatomical information and tissue characteristics, such as calcification, but also provide a more comprehensive assessment of patients with metastatic tumours.⁶ Magnetic resonance imaging can help to distinguish between benign and malignant tumours, it also can be used to determine the extent of myocardial, pericardial, and coronary artery invasion as well as provide more anatomical

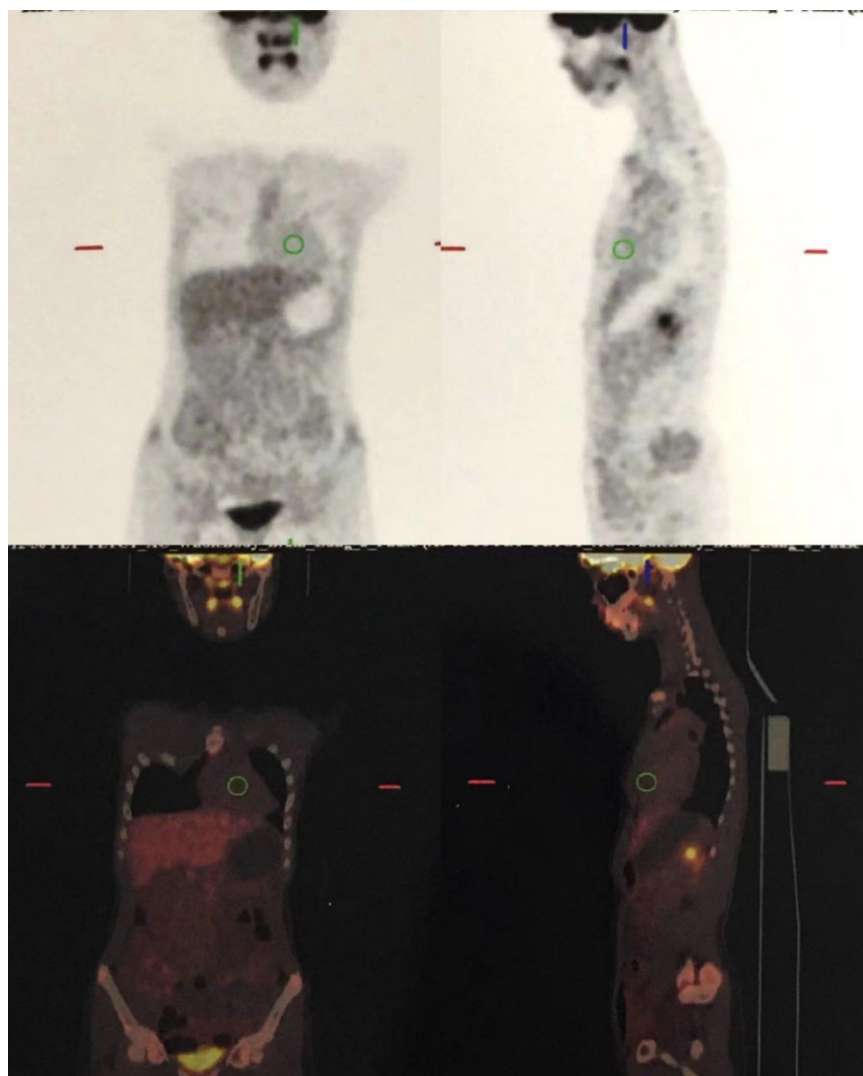


Figure 4 The appearance of the lesion on PET-CT. There was no increase in glucose metabolism was observed.

information for the operation. However, the imaging findings of some tumours are not specific, and the benign and malignant lesions are difficult to determine. At this time, PET-CT is needed for auxiliary diagnosis. Studies have shown that fluorodeoxyglucose-PET may be particularly suitable for distinguishing malignant and benign heart disease and assessing the recurrence of intracardiac tumours after resection.⁷ Therefore, it is important to make use of a comprehensive multi-modal evaluation and diagnosis before surgery, which can provide imaging support for the formulation of a surgical plan.

Fibromas need to be differentiated from other tumours of the heart, such as rhabdomyomas. Rhabdomyoma is the most common primary cardiac tumour in infants, accounting for more than 60% of cases. Rhabdomyoma present as a hyperechoic solid mass in echocardiography. On MRI, however, they enhance as isointense or slightly hyperintense on T₁WI and hyperintense on T₂WI. The enhancement degree of rhabdomyoma is less than that of myocardium on enhanced scans.⁸ Fibroma must also be differentiated from myxoma. Myxomas

are the most common primary cardiac tumours in adults, and most often occur in the left atrium. On ultrasound scans, myxoma present as a hyperechoic mass with a pedicle that shifts during the cardiac cycle. Myxoma present as isointense on T₁W1, hyperintense on T₂W1, and show obvious enhancement with contrast material.⁸ Another differential consideration for fibromas are metastases. They can invade the pericardium, the myocardium, and the intracardiac cavity.⁹ Most of these tumours are nodular in shape and size. The ultrasonic manifestations of metastases are masses in the cardiac cavity or pericardium, an uneven thickening of the ventricular wall, and uneven pericardium or pericardial effusion.¹⁰

When fibroma patients do not present with related clinical symptoms, it may not be treated, and the patient will be followed up for observation. Although the patient had no clinical symptoms, she had a heart murmur on auscultation, and the electrocardiogram showed arrhythmia, so she was treated with surgery. Complete resection is the primary choice of treatment for symptomatic fibroma.¹¹

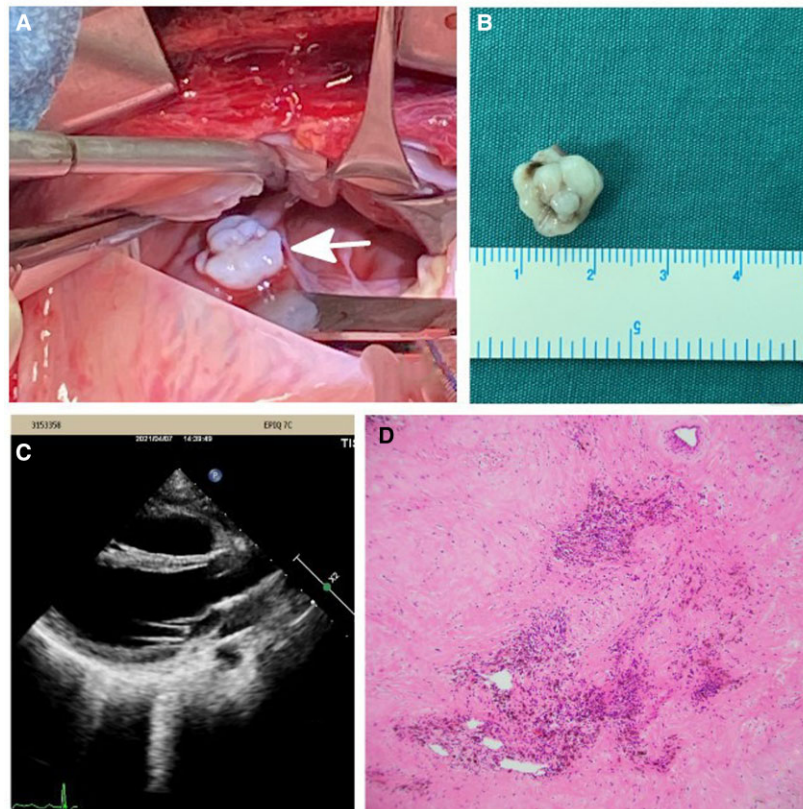


Figure 5 (A) During the operation, the tumour was located in the right ventricular septum, it was milky white, and its pedicle was located near the apex of the ventricular septum (arrow); (B) General picture of the tumour cut off during the operation; (C) Postoperative ultrasound showed no obvious abnormal mass echo; (D) Pathologically confirmed that this tumour is a fibroma.

Conclusions

Right ventricular fibroma is a rare primary cardiac tumour. Pathology is the gold standard for the diagnosis of fibroma. However, in the absence of pathology, it is necessary to use multi-modal imaging to evaluate the lesions and distinguish between benign and malignant tumours. Multi-modality imaging can help to identify the nature of cardiac tumours and can provide effective information for both preoperative and postoperative evaluation of patients.

Lead author biography



Pengtao Sun received a Bachelor of Medical Imaging from the Sun Yat-sen University. After graduating from the University of Sun Yat-sen, he transferred to Guangzhou University of Chinese Medicine and obtained his Master of Medicine. Currently, he is working as a Ultrasound doctor at Guangdong Provincial Hospital of Chinese Medicine in Guangzhou. His fields of

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Supplementary material

Supplementary material is available at *European Heart Journal – Case Reports* online.

Slide sets: A fully edited slide set detailing these cases and suitable for local presentation is available online as Supplementary data.

Consent: The authors confirm that written consent for submission and publication of this case series including images and associated text has been obtained from the patient in line with COPE guidance.

Conflict of interest: We declare that we have no financial and personal relationships with other people or organizations that can inappropriately influence our work, there is no professional or other personal interest of any nature or kind in any product, service and/or company that could be construed as influencing the position presented in, or the review of.

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