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Case Report

Preoperative angioembolisation of a mediastinal accessory ectopic spleen: A case report and review of the literature^{☆☆☆}

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ARTICLE INFO

Article history:

Received 7 March 2022

Revised 14 April 2022

Accepted 15 April 2022

Keywords:

Accessory spleen

Congenital variant

Interventional radiology

Diagnostic radiology

Mediastinal mass

Angioembolization

ABSTRACT

A 50-year-old male presented to our institution for embolization of an incidentally detected mediastinal mass prior to surgical resection. The patient had undergone extensive pre-procedural imaging as well as bronchoscopy and mediastinoscopy. Ultimately, resection was required for a definitive diagnosis of congenital ectopic mediastinal accessory spleen. This case represents the first reported incidence of ectopic splenic tissue in this location and illustrates the difficulties in establishing a pre-operative diagnosis with often confounding imaging findings.

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Introduction

Development of the spleen begins during the 5th week of life, arising from the intraembryonic splanchnic mesoderm [1]. Pre-splenic tissue is initially located in symmetric domains

on both sides of the embryo with right sided tissue regressing during development [2]. Rotation of the stomach and development of the dorsal mesogastrium during the 6th and 7th weeks of life leads to the migration and fusion of multiple splenic buds into the left upper abdomen [3].

* Acknowledgments: The authors wish to thank Dr. Lee Bornman at Capital Pathology for his assistance in providing histopathology images.

☆☆ Declaration of Competing Interests: The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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<https://doi.org/10.1016/j.radcr.2022.04.030>

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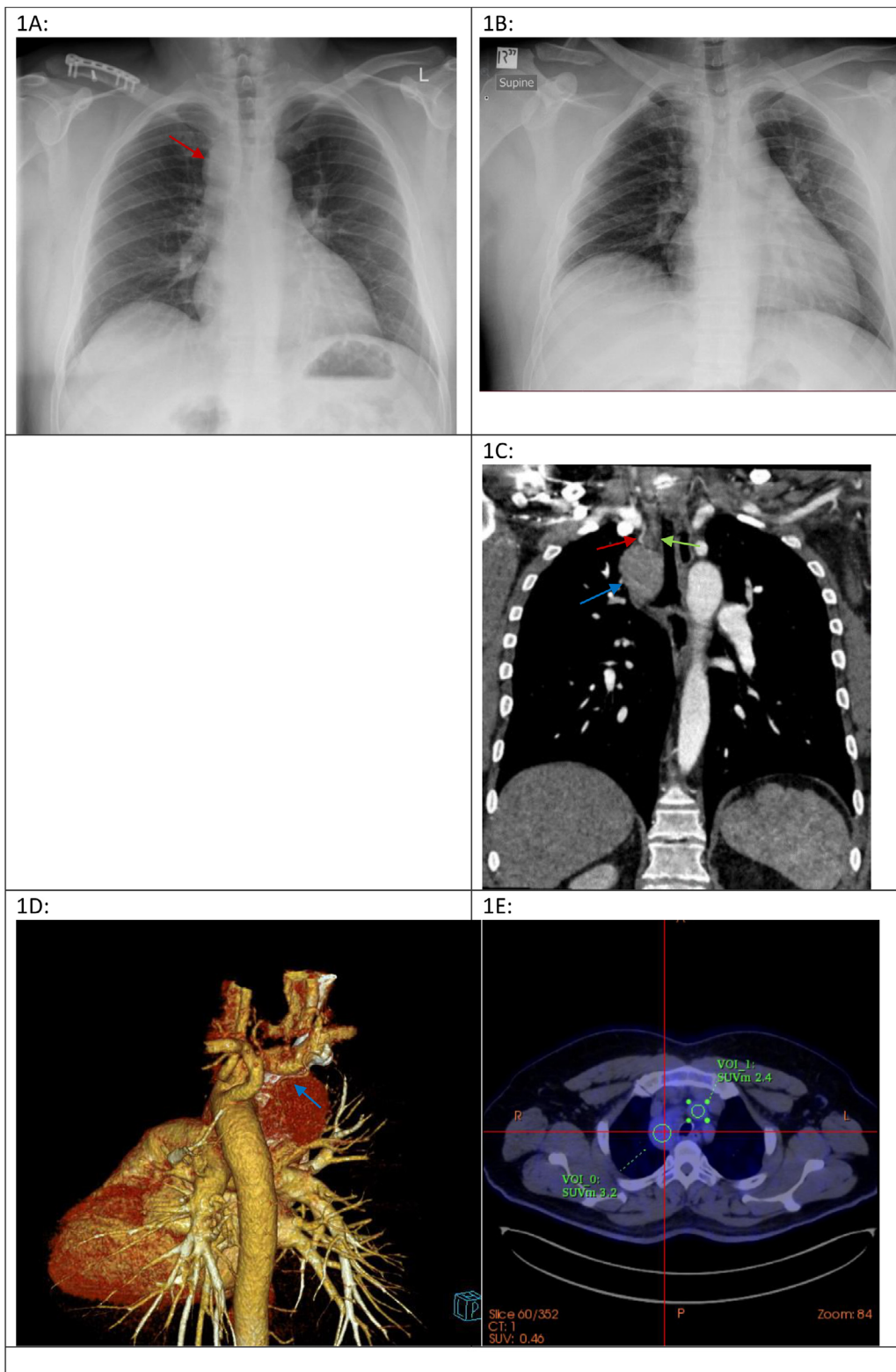


Fig. 1 – (A) PA Chest radiograph demonstrating the right paratracheal mass with subtle tracheal effacement (arrow). **(B)** Supine trauma chest radiograph with subtle right paratracheal mass. **(C)** Coronal CT MIP demonstrating a feeding artery from right subclavian (red arrow), paratracheal mass (blue arrow), satellite isodense nodule (green arrow), and native spleen. Note isodense enhancement. **(D)** posterior oblique 3D reconstruction demonstrating anomalous branch supplying the hypervascular mass (blue arrow) **(E)** Axial CT PET demonstrating mild avidity of the mass when compared to the blood pool. (Color version of figure is available online.)

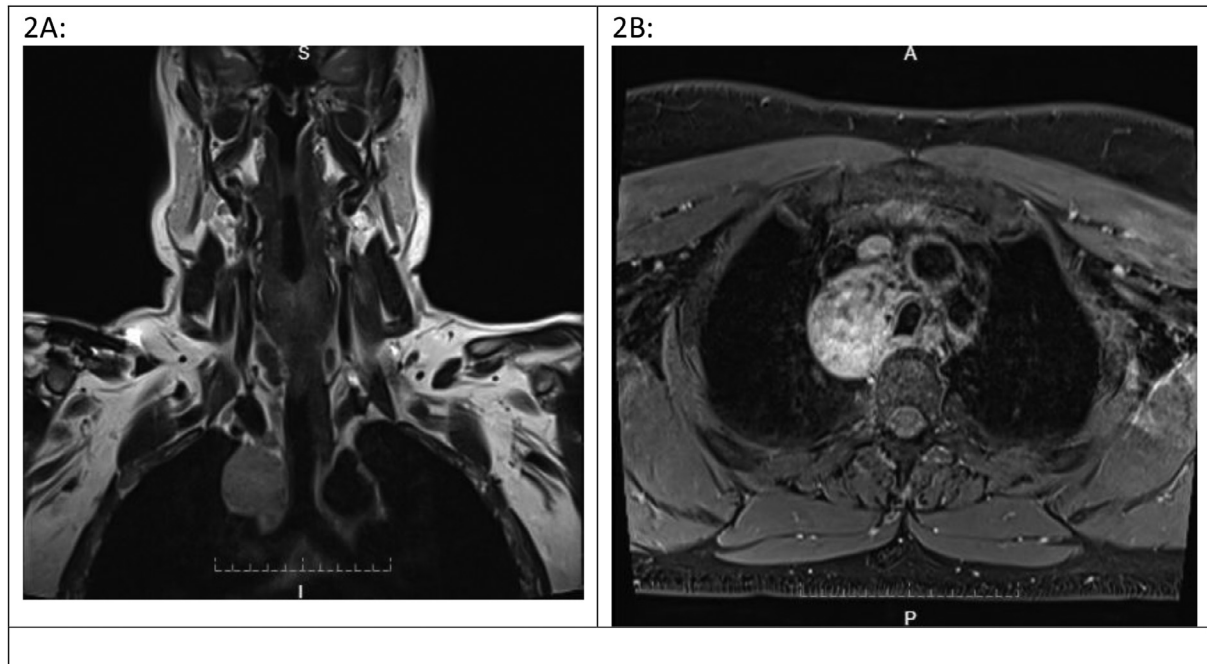


Fig. 2 – (A) Coronal T1 MRI showing hyperintense mass. (B) Axial T1 fat saturated post gadolinium contrast image demonstrating enhancement

Congenital accessory splenic tissue is a relatively common occurrence with an incidence at autopsy between 10% and 30%. It is most commonly located within the perisplenic region and rarely measures larger than 40mm in maximal dimensions [4,5]. Less frequently, other locations have been reported including within the retroperitoneum, intrapancreatic, left intragonadal, and very rarely intrathoracic [6–11]. In the case of accessory intrathoracic splenic tissue, it has been postulated that aberrant migration into the pleural cavity may occur via incomplete pleuroperitoneal folds [8].

Case Report

A 50 year old male was referred for specialist investigation of a right paratracheal mass reported on chest X-ray (Fig. 1A) and CT performed for back pain.

Review of historical trauma series radiographs from 13 years prior revealed an unreported equivocal paratracheal opacity (Fig. 1B).

Prior to admission the patient had undergone multiple imaging studies including multiphase CT, CT PET, and an MRI of the mediastinum. Multiphase CT demonstrated a 44 mm AP x 35 mm transverse x 50 mm CC well marginated homogeneously enhancing mass with prominent surrounding vessels and a feeding artery emanating from the proximal right subclavian coursing between the mass and the trachea. There was mild mass effect on the trachea and no evidence of invasion of adjacent structures. A separate isodense 16 mm nodule was located immediately cephalad to the dominant mass (Figs. 1C and D). Of note, neither demonstrated the typical heteroge-

nous red pulp arterial phase physiologic enhancement, however, retrospective analysis of density revealed similar values (approximately 95HU) for both the lesion and the native spleen. This finding is concurrent with the findings of previous analyses, showing that the majority (77%) of accessory spleens are isodense when compared to their native counterparts [12]. Retrospective comparison with prior imaging suggested no change in size over a 3 month interval.

On CT PET imaging, the lesion demonstrated only mild avidity (SUV max 3.2 vs blood pool SUV max 2.3) (Fig. 1E).

Gadolinium enhanced MRI revealed a T1 isointense, T2 hyperintense ovoid mass with mild post contrast enhancement but no further distinguishing features (Figs. 2A and B). No further concerning masses or evidence of primary neoplastic disease were identified.

The patient underwent endobronchial ultrasound guided fine needle aspiration biopsy which revealed a mixed population of lymphocytes expressing both T and B cell antigens without any abnormal immunophenotypic features. Tracheal and bronchial washings were negative for any evidence of malignant cells.

The patient subsequently underwent mediastinoscopy, with the intention of biopsy, however, this was abandoned following visualization of multiple prominent peri-lesional vessels and contact bleeding. A decision to embolize the mass prior to resection was made, and the patient consented accordingly.

Past medical history included gastroesophageal reflux, prior clavicular fracture and fixation, and lumbar disc degeneration. Of note, there was no history of abdominal trauma or diaphragmatic injury.

The patient's only regular medication was Pantoprazole 40 mg once daily.

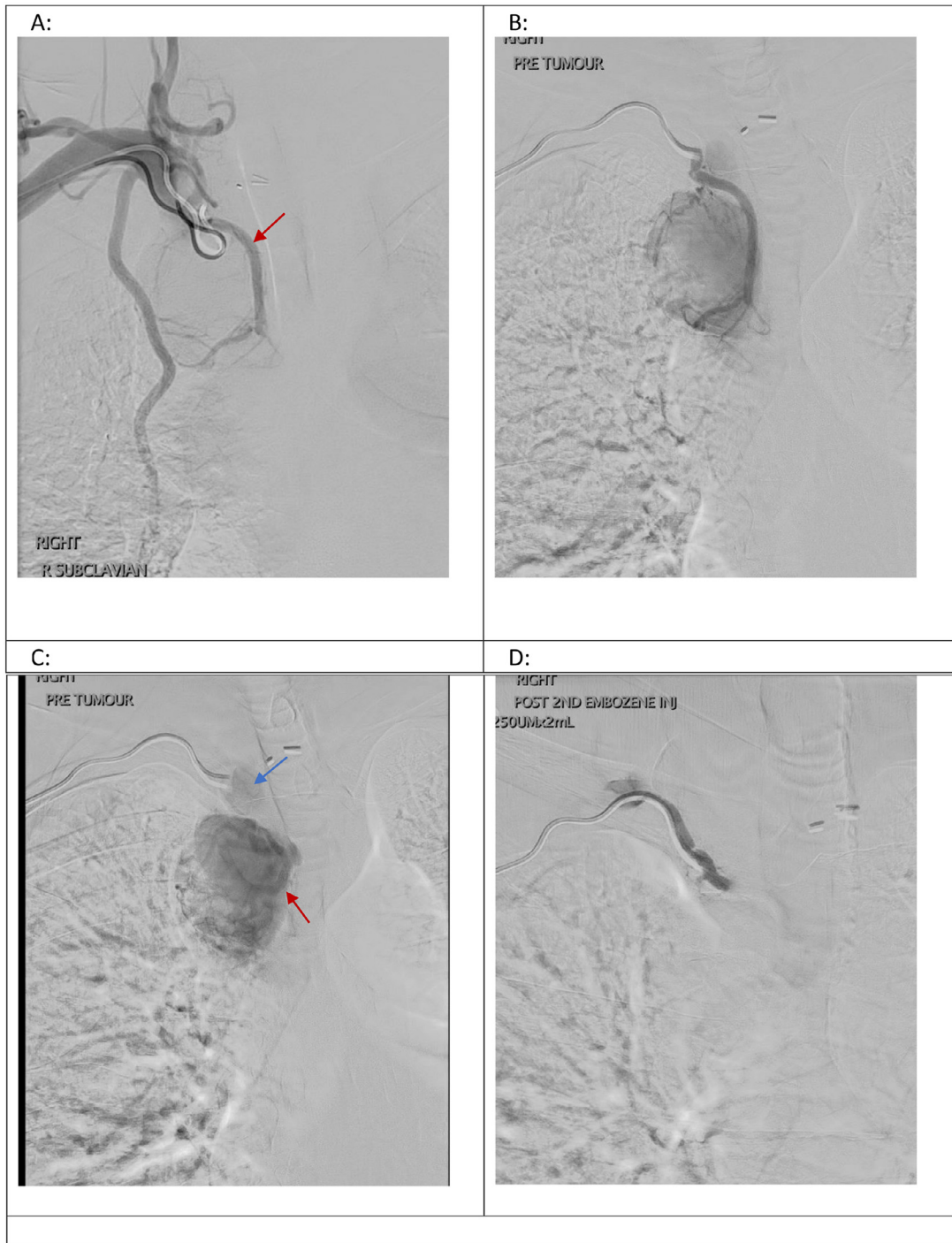


Fig. 3 – (A) Right subclavian arteriogram demonstrating anomalous branch (red arrow) supplying the paratracheal mass. (B) Superselective injection showing vascularity. (C) Prominent draining veins (red arrow) and satellite nodule (blue arrow). (D) Post-embolization control shows complete devascularization. (Color version of figure is available online.)

Potential differential diagnoses which were raised during the clinical workup included hemangioma, paraganglioma, Castleman's disease, and lymphoma.

Bloodwork was unremarkable with normal full blood count, UECs, LFTs, and Coagulation profile. Catecholamines and tumor markers were negative.

Right radial arterial access was obtained under ultrasound guidance and a 4Fr sheath inserted. Digital subtraction angiography demonstrated an arterial supply to the mass arising from a dedicated branch of the right subclavian artery. Small draining veins were also present. Super selective embolization of the feeding artery was accomplished using a 2.4Fr ProGreat

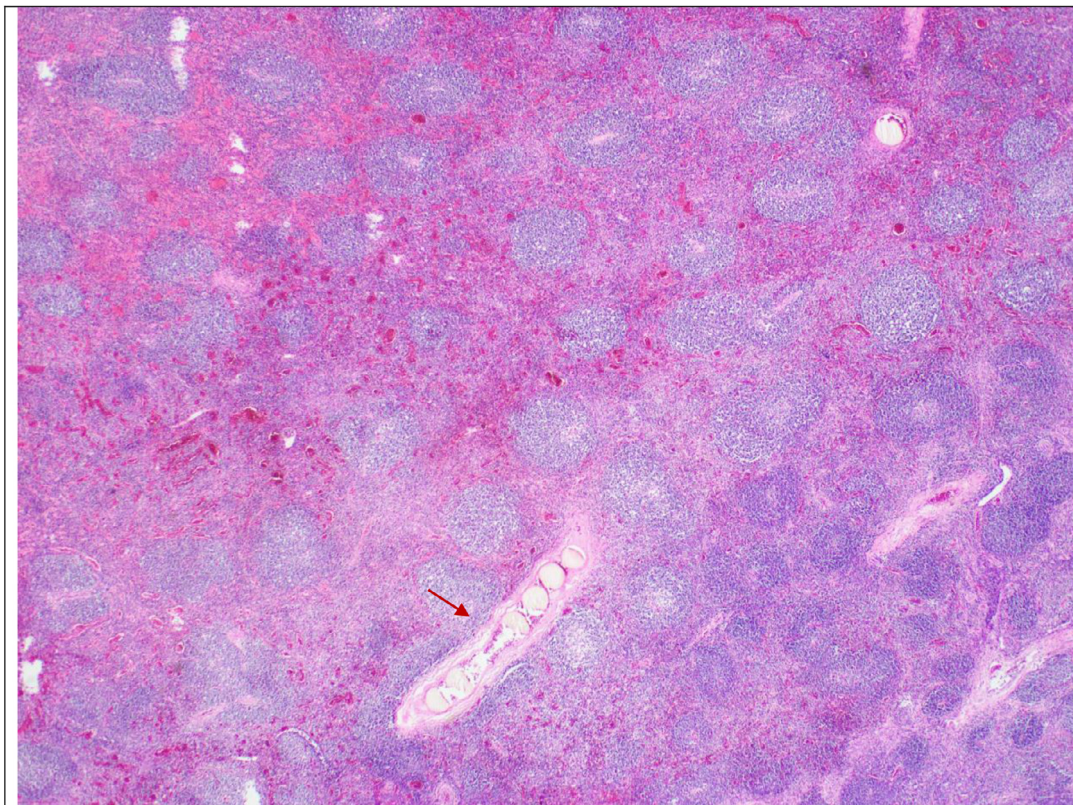


Fig. 4 – Low power H&E stain demonstrating classic histological features of splenic tissue including red and white pulp. Yellow Embolus particles are visible in several small vessels (arrow). (Color version of figure is available online.)

Microcatheter (Terumo Corporation, Tokyo, Japan) supported by a 4Fr Mikaelson catheter (Cook Medical, Bloomington, IN, USA) and 250um Embolus particles (Varian Medical Systems, Palo Alto, CA, USA) followed by a single VortX 5×4.5mm fibred coil (Boston Scientific, Boston, MA, USA) (Figs. 3A and D).

24 hours post embolization, the patient underwent a right thoracotomy and en-bloc resection of the devascularized mass with minimal blood loss and made an uneventful post-op recovery.

Both intraoperative frozen sections and final histopathology revealed tissue composed of white pulp areas with numerous lymphocytes forming follicles composed of mixed B and T cell populations (Fig. 4). CD3, CD20, CD21, CD8, CD34, and CD68 immunohistochemical stains further demonstrated splenic type architecture. Despite effective devascularization, there were no histologic features of infarction. Of note, CK5 staining was negative, excluding a thymic lesion. Flow cytometry did not reveal any evidence of non-Hodgkin Lymphoma and a final diagnosis of accessory ectopic spleen was established.

Discussion

Congenital ectopic thoracic spleen represents an exceedingly rare clinical entity with only 5 cases reported in the literature [6–9,11]. Four of these case reports described tissue within the

left hemithorax [6,7,9,11] and 1 within the right hemithorax [8]. To our knowledge, this case represents the first reported incidence of congenital accessory splenic tissue within the middle mediastinum. The authors postulate this rare entity may be secondary to failure of regression of right sided embryological splenic tissue and/or errors in migration.

This case illustrates the difficulty in diagnosis of such a rare and previously unreported entity. Multiple imaging tests and modalities were insufficient and ultimately, in line with previously reported cases, the patient required surgery for a definitive diagnosis [6–9,11]. The use of denatured red blood cell or Sulphur colloid radionuclide studies was not entertained in our patient, but could have proved diagnostic as is the case in splenosis.

It is hoped this case report will raise awareness of this extremely rare condition, potentially aiding clinicians in future when assessing patients with stable and non-aggressive appearing mediastinal masses.

Patient Consent

Informed and written consent was obtained from the patient for the purposes of publication of this case report. The patient signed the Elsevier Patient Consent Statement. This document has been retained by the department and can be provided on request.

Supplementary materials

Supplementary material associated with this article can be found, in the online version, at doi:10.1016/j.radcr.2022.04.030.

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