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A Rare Case of Pediatric Ventricular Pseudoaneurysm as a Sequelae of Ascending Aortic Dissection

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Abstract: ***Background**: Aortic dissection is a critical cardiovascular condition characterized by the separation of the aortic wall layers, leading to the formation of a false lumen that can compromise blood flow and result in severe complications. While it is predominantly observed in adults, pediatric cases, although rare, can occur and may lead to unusual sequelae such as ventricular pseudoaneurysms. **Case presentation**: A 9 - year - old boy with complaints of breathlessness for a month and vague chronic chest pain. ECG and ECHO were performed which showed chronic ischemic changes and inferior wall aneurysm. On CT examination, the topogram revealed a well- defined elliptical diffuse eggshell calcification involving most of the left cardiac silhouette. CT thoracic angiogram showed a well - defined elliptical outpouching arising from the inferior wall of the left ventricle with a thin rim of wall calcification. A peripheral wedge - shaped non - enhancing area in the lower pole of the spleen was seen, suggesting a splenic infarct. **Conclusion**: This case report illustrates a rare but significant sequela of ascending aortic dissection in a pediatric patient—a ventricular pseudoaneurysm. It serves as a reminder of the complexities involved in diagnosing and managing aortic dissections in younger populations.*

Keywords: Aortic dissection, Ventricular pseudoaneurysm, Pediatric cardiac disease

1. Introduction

Aortic dissection is a critical cardiovascular condition characterized by the separation of the aortic wall layers, leading to the formation of a false lumen that can compromise blood flow and result in severe complications. While it is predominantly observed in adults, pediatric cases, although rare, can occur and may lead to unusual sequelae such as ventricular pseudoaneurysms. This manuscript presents a case report of a pediatric patient who developed a ventricular pseudoaneurysm following ascending aortic dissection, emphasizing the importance of timely diagnosis and management.

2. Case Presentation

A 9 - year - old boy with complaints of breathlessness for a month and vague chronic chest pain. ECG and ECHO showed findings of chronic ischemic changes and an inferior wall aneurysm. On CT examination, the topogram (Fig.1) revealed a well - defined, elliptical, diffuse eggshell calcification of size 8.1 x 5.2 cm involving most of the left cardiac silhouette.

CT thoracic angiogram (Fig.2) showed a well - defined, elliptical outpouching arising from the inferior wall of the left ventricle with a thin rim of wall calcification. The lesion was seen within the pericardial cavity causing displacement of the cardiac apex superiorly and left dome of the diaphragm inferiorly. Contrast enhancement (Fig.3) was similar to the lumen of the left ventricle in all phases. The neck of the lesion measured 2.3 x 2.0 cm (narrow). The base of the lesion showed a focal, non - calcified, dehiscent wall with intact pericardium. The ascending aorta showed a thin dissection from the aortic root to the proximal arch. Left ventricular hypertrophy was also visualized.

Left basal lobe consolidation with mild pleural effusion was noted. A peripheral wedge - shaped non - enhancing area in the lower pole of the spleen was seen, suggesting a splenic infarct.

The diagnoses of an ascending aortic dissection [Stanford type B], calcified discrete saccular left ventricular pseudoaneurysm with features of left ventricular failure [on echo correlation], and a splenic infarct were made.

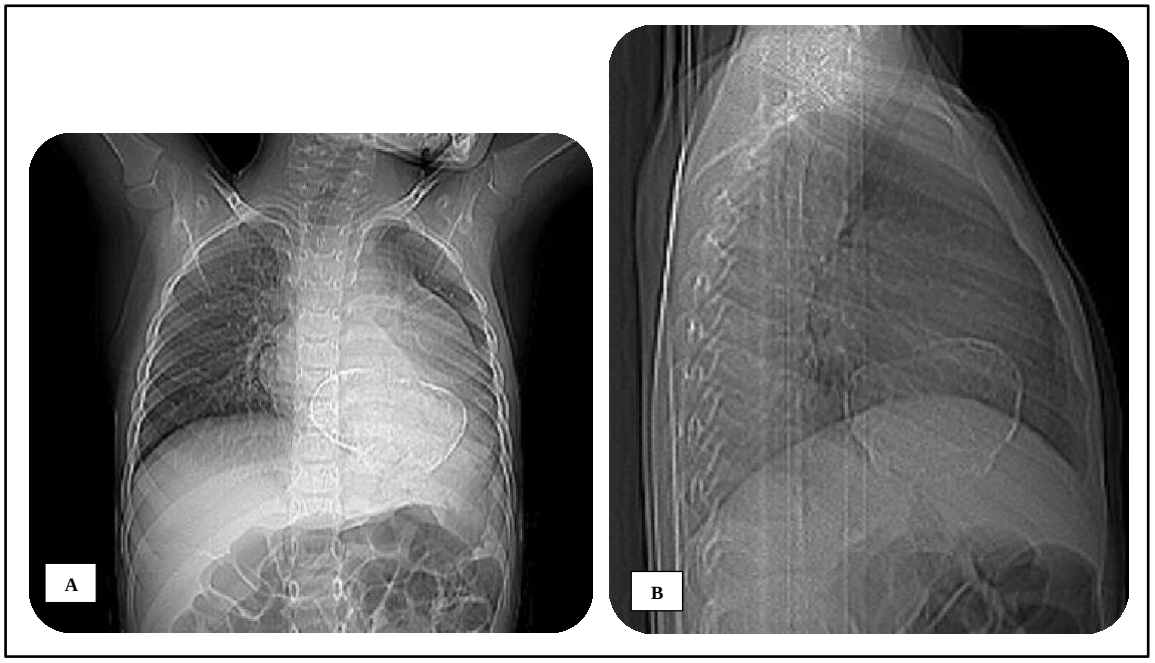

Figure 1 (A - B): CT topogram shows a well - defined, elliptical, diffuse eggshell calcification of size 8.1 x 5.2 cm, involving most of the left cardiac silhouette.

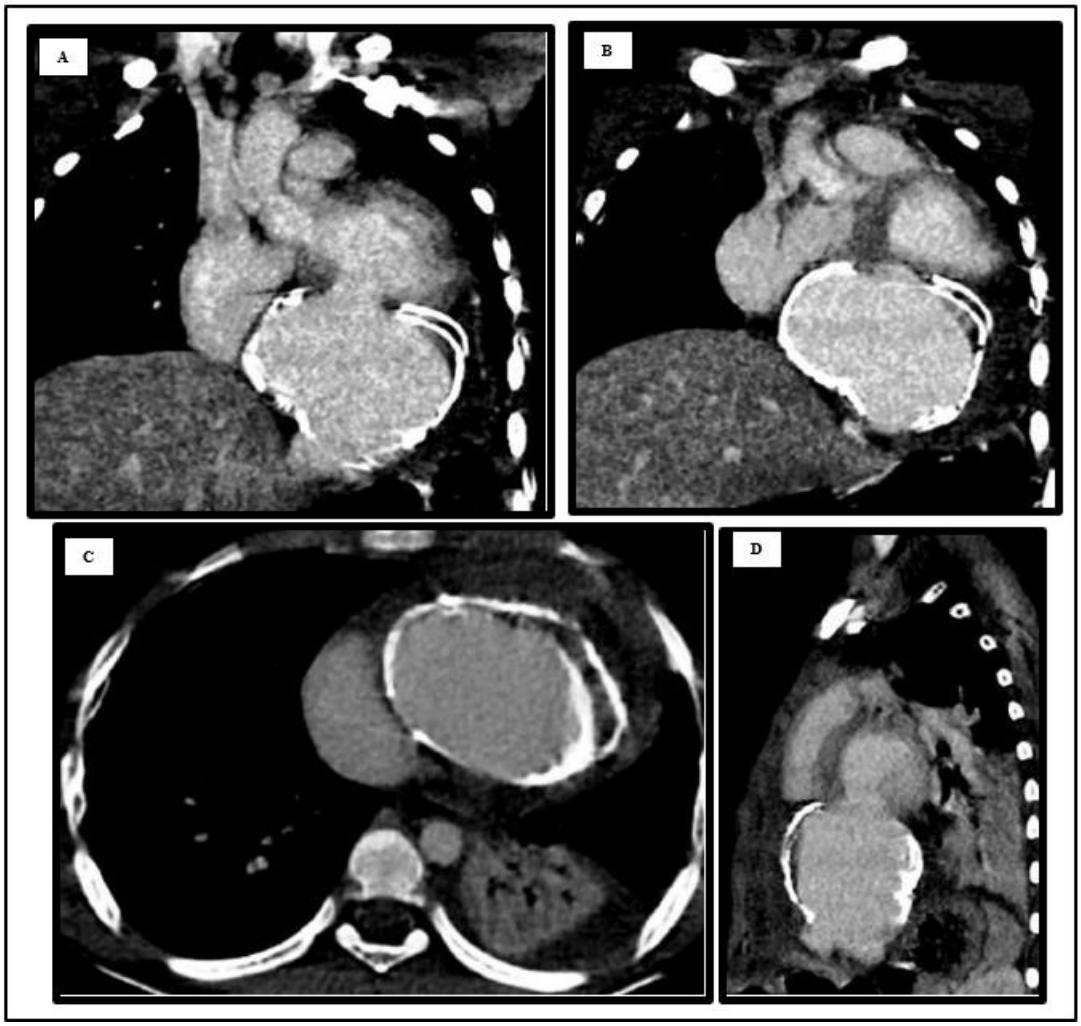

Figure 2 (A - D) - CT thoracic angiogram shows a well - defined, elliptical outpouching arising from the inferior wall of the left ventricle showing a thin rim of wall calcification. The lesion is within the pericardial cavity causing displacement of the cardiac apex superiorly and the left dome of the diaphragm inferiorly. The neck of the lesion appears narrow measuring 2.3 x 2.0 cm. The base of the lesion shows a focal non - calcified dehiscent wall with intact pericardium.

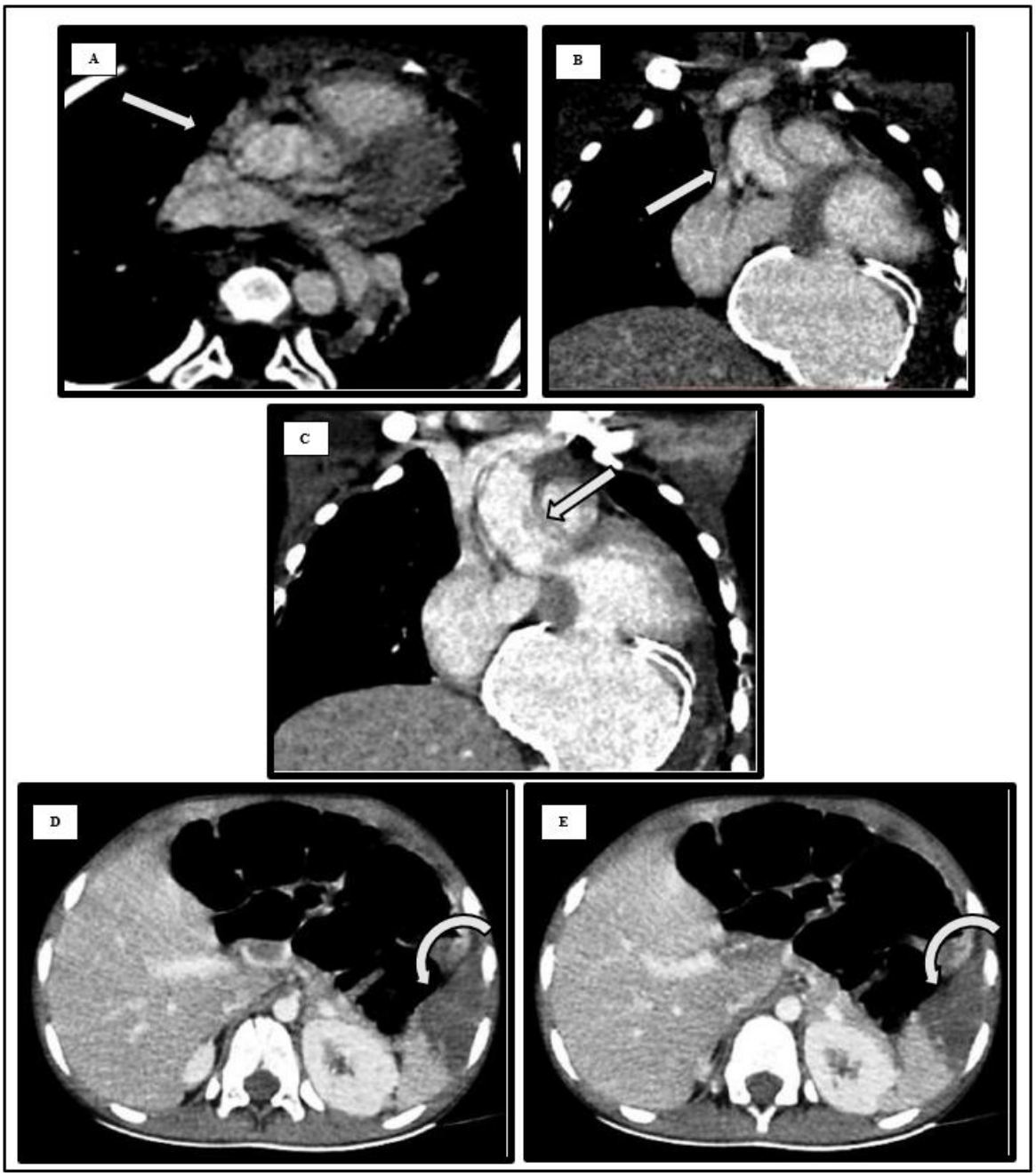

Figure 3 (A - E): Contrast - enhanced images show the enhancement of the outpouching to be similar to the lumen of the left ventricle in all phases. The base of the lesion shows a focal non - calcified dehiscent wall with intact pericardium. The ascending aorta shows a thin dissection from the aortic root to the proximal arch (straight arrow). Left ventricular hypertrophy is seen. Left basal lobe consolidation with mild pleural effusion seen. A peripheral, wedge - shaped, non - enhancing area in the lower pole of the spleen (curved arrow) is seen, suggesting a splenic infarct.

3. Discussion

Ventricular aneurysms are rare in children. It can be either a true aneurysm or a pseudoaneurysm. Ventricular pseudoaneurysm is a free wall rupture of the left ventricle contained by the pericardial tissue adjacent to it[1]. True aneurysms are mostly congenital[2, 3].

Left ventricular pseudoaneurysms form when a cardiac rupture is contained by adherent pericardium or scar tissue and theories regarding causative factors vary[4]. Spontaneous focal pseudoaneurysm due to focal abnormality in the medial layer; may occur in normal caliber or aneurysmal aorta[5]. Associated with trauma, infection, an anomalous left coronary artery from the pulmonary artery, vasculitis, intrauterine myocardial ischemia, and surgical intervention[6, 7].

Aortic dissection in children is often associated with underlying connective tissue disorders, such as Marfan syndrome or Ehlers - Danlos syndrome, which predispose them to vascular abnormalities[8, 9]. Its incidence ranges between 5 to 30 cases in a million people per year[10].

Aortic dissection can be classified using either of two classifications based on anatomical location, Stanford and DeBakey classification. Stanford type A dissections are present in the ascending aorta and type B dissections are

seen distal to the subclavian artery[10]. Aortic dissection with extension to coronary arteries leading to ischemia complicating as ventricular aneurysm is a rare presentation which is depicted in our case.

In this case report, the patient presented with symptoms consistent with acute aortic dissection, including severe chest pain and hemodynamic instability. The diagnosis was confirmed through imaging modalities, primarily contrast - enhanced computed tomography (CECT), which is the gold standard for identifying the extent and nature of the dissection[5, 10].

Chronic heart failure, chest pain, and dyspnea are the most frequently reported symptoms. Arterial embolization (most commonly: splenic infarct, as in our case) and arrhythmias have also been described, while >10% of patients are asymptomatic[11].

The presence of a ventricular pseudoaneurysm as a complication of ascending aortic dissection is particularly concerning. This condition arises when blood leaks from the aorta into the myocardium, creating a false chamber that can rupture if not promptly addressed[12]. The management of such complications typically involves a surgical intervention to repair both the dissection and the resultant pseudoaneurysm. In this case, the patient underwent successful surgical repair, highlighting the critical role of early intervention in preventing life - threatening outcomes[9, 10].

Echo, CT, and MRI aid in the diagnosis of ventricular pseudo aneurysm and differentiate it from a true aneurysm. Distinguishing between these two types of aneurysm (Table) is important because treatments differ[7, 13]. True aneurysms have a low likelihood of complications and are generally treated through elective surgery or even conservatively whereas pseudoaneurysms have a high rate (45%) of rupture and need urgent surgical intervention[14].

Recent literature underscores the need for heightened awareness among clinicians regarding atypical presentations of aortic dissection in pediatric patients. The differential diagnosis should include considerations for myocardial ischemia and other cardiac complications that may mimic or coexist with dissection. Furthermore, guidelines from organizations such as the American Heart Association emphasize the importance of utilizing risk stratification tools like the Aortic Dissection Detection Risk Score (ADD - RS) to facilitate timely diagnosis.

4. Conclusion

This case report illustrates a rare but significant sequela of ascending aortic dissection in a pediatric patient—a ventricular pseudoaneurysm. It serves as a reminder of the complexities involved in diagnosing and managing aortic dissections in younger populations. Clinicians must maintain a high index of suspicion for such complications and utilize appropriate imaging techniques for accurate diagnosis. Early surgical intervention remains paramount in mitigating the risks associated with this life - threatening condition.

List of Abbreviations:
ECG - Electrocardiography ECHO - Echocardiography
CT - Computerised Tomography
MRI – Magnetic Resonance Imaging CE – Contrast Enhanced

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