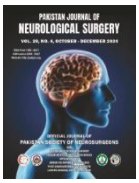

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Case report

Incidental Findings of Dextrocardia in Patient with Cerebellar Arteriovenous Malformation

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ABSTRACT

Cerebral arteriovenous malformations (AVMs) have a congenital origin, and the diagnosis rate during infancy and youth is only 18-20%. The clinical presentation of intracranial haemorrhage is observed in 75-80% of paediatric patients. The introduction of novel agents for endovascular management has led to enhanced surgical outcomes by preoperative AVM embolization. Dextrocardia with situs inversus is also a rare congenital abnormality in children. A significant proportion of individuals diagnosed with dextrocardia remain oblivious to their condition due to the prevalence of asymptomatic cases. We are documenting a case involving a 12-year-old girl who was presented with an altered state of consciousness and vomiting. Her Neuroimaging showed posterior fossa bleed and diagnosed with cerebellar arteriovenous malformation (AVM). Endovascular embolization of AVM was done and an incidental finding of dextrocardia with situs inversus was noted during the procedure. This case report aims to find if there is any association between Brain AVMs and Dextrocardia in the paediatric population.

Keywords: Dextrocardia, Arteriovenous Malformation.

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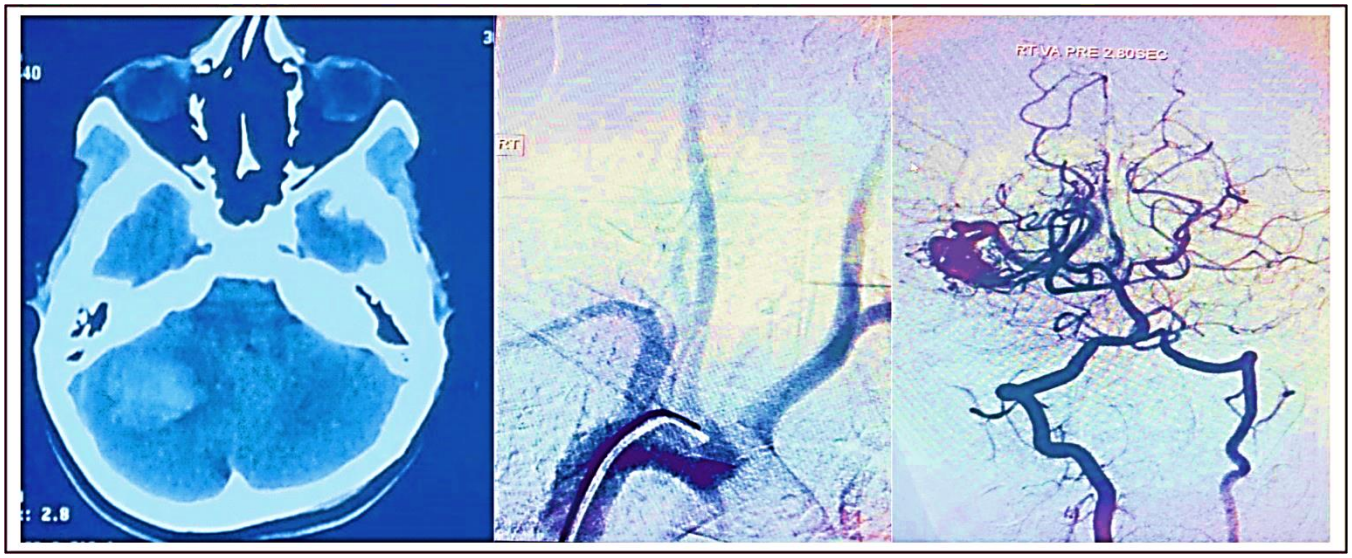
INTRODUCTION

Cerebral parenchymal arteriovenous malformations (AVMs) are predominantly identified between the age range of 20 to 40 years, with a mere 18-20% of cases manifesting symptoms before the age of 15.[1-2] Haemorrhage is a prevalent clinical manifestation observed in both adults and children, with a higher incidence rate observed in the paediatric population compared to the adult population (75-80% versus 50-65%).[3] Dextrocardia is also a rare congenital anomaly marked by the heart being repositioned to the right side of the thoracic cavity, often accompanied by the inverse rotation of all internal organs (known as situs inversus totalis).[4-5] The occurrence of this anomaly is rare, with an incidence rate of 1 in 10,000 live births.[6] Dextrocardia sometimes goes unnoticed by

individuals until they seek medical assistance for unrelated diseases.[7] The presence of congenital cardiac abnormalities and other deformities may be observed in individuals diagnosed with dextrocardia and situs inversus.[8] In this study, we document an atypical instance of dextrocardia with situs inversus and Cerebellar AVM in a young patient admitted with posterior fossa intracranial haemorrhage.

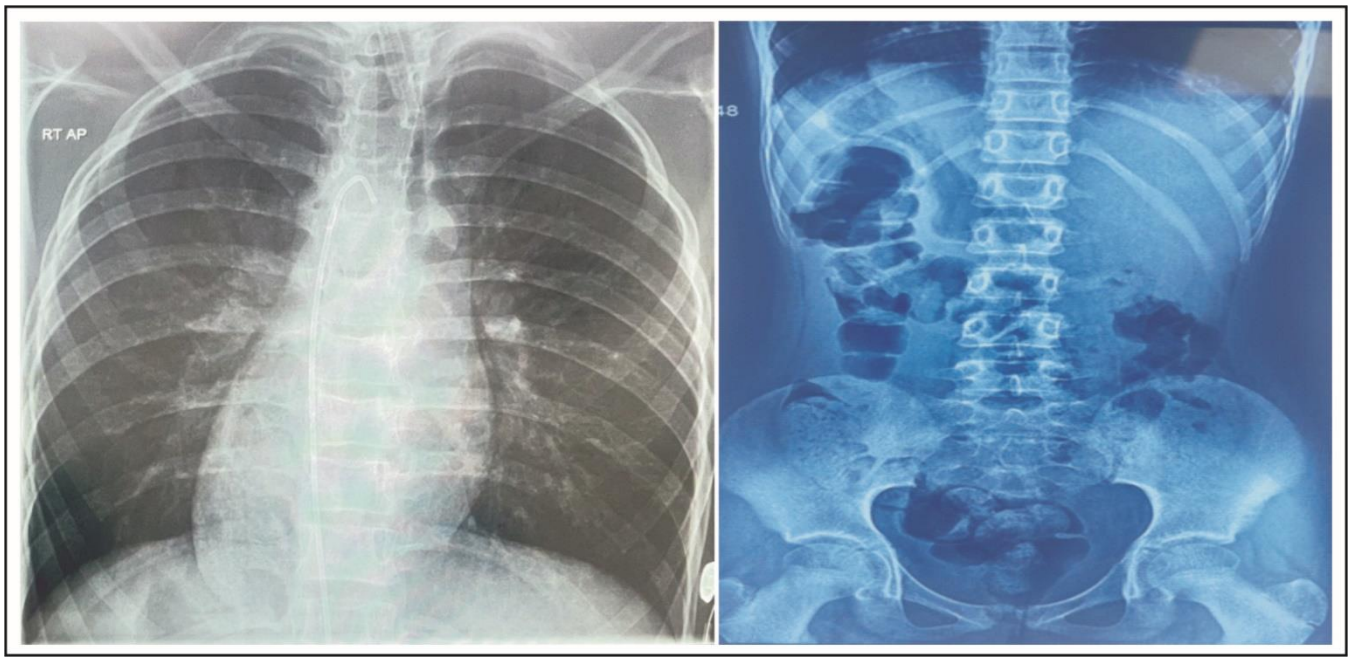
CASE REPORT

Twelve-year-old girl presented to the emergency department with complaints of sudden severe headache and multiple episodes of vomiting followed by altered sensorium in February 2024 and was admitted to Punjab Institute of Neurosciences Lahore. The patient did not have any history of fits or cardiopulmonary symptoms including nasal congestion, chronic coughs, palpitations or any other metabolic disorders. She had no history of any hospital admission before. On neurological examination, her muscle tone was slightly increased. A physical exam of her cardiovascular system discovered a pulse rate of 105 beats/min and her blood pressure was 100/60 mmHg. Additionally, her lungs were clear on auscultation. Her routine labs were normal. CT brain plain showed a right cerebellar haemorrhage (Figure 1A). A provisional diagnosis of cerebellar AVM was made and the Cerebral DSA and embolization procedure was planned. After aseptic measures, under general anaesthesia, 6fr femoral sheath was placed and 5fr hink diagnostic catheter advanced to the aortic arch over the guide wire, Arch angiography demonstrated dextrocardia (Figure 1 B) and post-procedure chest X-ray confirmed the same. (Figure 2 A) and aortic arch contrast runs (Figure 1 B). Bilateral ICA and Right VA angiography Intracranial runs were taken (Figure 1, C). Embolization of AVM was performed with Onyx 18 through the Right SCA feeder and post embolization run was taken. No Post embolization deficit or complication was noted. Post procedure Echocardiography findings (Table 1) and X-ray abdomen erect were done to confirm dextrocardia and situs inversus (Figure 2 B). The patient was discharged with no focal deficit. On a follow-up visit after 2 weeks and 1 month, the patient was asymptomatic with no deficit.



A) **B)** **C)**

Figure 1: CT Brain Plain showing right cerebellar ICH. **B)** Showing Arch and large vessels **A)** DSA Right VA AP View Pre-Embolization (Images added with patient consent).



A)
Figure 2A): Chest X-ray showing dextrocardia.

B)
B) X-ray Abdomen Erect (Images added with patient consent).

Table 1: Echocardiography Findings.

Imaging Modality	Findings
Echocardiography	Dextrocardia with Situs inversus. Otherwise, Normal resting cardiac echo study.

DISCUSSION

The prevalence of Cerebral arteriovenous malformations (AVMs) in pediatric patients is 0.014±0.028%. The proportion of cases identified under the age of twenty ranges from 19.6% to 42%. Paediatric patients had a higher annual risk of haemorrhage (3.2%) compared to adults (2.2%). The incidence of rebleeding is 6.33% in the initial year following the first haemorrhage and subsequently decreases, reaching a stable level during the fourth year.[1] This patient presented with an Intracranial haemorrhage. The patient had a history of headaches as well. While diagnosing the aetiology of bleed and treating the cerebellar AVM, the patient was diagnosed with dextrocardia incidentally. The aetiology of dextrocardia remains unknown; however, it has been suggested that maternal diabetes mellitus and maternal cocaine usage may be contributing factors. There is also a suspicion of genetic factors, as evidenced by a higher occurrence observed in conjoined twins. The concept of situs inversus totalis does not exhibit any sex or racial predilection.[9] Dextrocardia with situs inversus presents a significant risk due to its asymptomatic nature and tendency to go misdiagnosed unless it was accidentally identified during an investigation for another medical condition. The alignment of visceral and brain asymmetry observed in some vertebrate species raises the question of whether this association also exists in humans. While the visceral and brain systems may have developed asymmetry for different reasons, basic visceral left-right differentiation mechanisms could have been duplicated to establish brain asymmetry.[10] Dextrocardia associated with situs inversus and

other cyanotic and acyanotic congenital heart diseases can present with brain manifestations like altered hemodynamics increased intracranial tension and brain abscess. Complicated cardiac malformation like this when associated with brain abscess poses several challenges.[11] But in our case, there was no other associated cardiac anomaly noted on further cardiac workup. The presence of dextrocardia with situs inversus is usually an incidental report and may not result in any symptoms or health complications. Nevertheless, the correlation between cerebral AVMs and this condition is not firmly established and could perhaps be a fortuitous event in this particular instance. This case report will document these findings which may help to identify the association between brain AVMs and Dextrocardia and their relation with each other due to haemodynamic changes for larger studies in future.

CONCLUSION

No significant association between Brain AVMs and Dextrocardia has been reported in the literature. There may be haemodynamic abnormality associated with dextrocardia which can cause brain AVMs. This particular case highlights the significance of systematic evaluation and management of paediatric patients presenting with neurological symptoms, as underlying pathologies such as cerebral AVMs may require prompt intervention to prevent serious complications. Additionally, the incidental finding of dextrocardia underscores the need for a comprehensive assessment to detect any associated anomalies or conditions, even if unrelated to the primary complaint.

Informed Consent: Written informed consent was obtained from the patient for case report publication.

Conflict of Interest & Grant Support: No competing interests.

Disclosure Statement: The authors declare that they have no relevant or material financial interests that relate to the research described in this paper.

REFERENCES

1. Millar C, Bissonnette B, Humphreys RP. Cerebral arteriovenous malformations in children. Canadian journal of anaesthesia. 1994;41:321-31. Doi: 10.1007/BF03009913
2. Humphreys R. Arteriovenous malformations of the brain. Concepts Pediatr Neurosurg. 1988;8:146-64. Available from: cir.nii.ac.jp/crid/1574231874473362560
3. Jomin M, Lesoin F, Lozes G. Prognosis for arteriovenous malformations of the brain in adults based on 150 cases. Surgical neurology. 1985;23(4):362-6. Doi: 10.1016/0090-3019(85)90207-1
4. Bohun CM, Potts JE, Casey BM, Sandor GG. A population-based study of cardiac malformations and outcomes associated with dextrocardia. The American journal of cardiology. 2007;100(2):305-9. Doi: 10.1016/j.amjcard.2007.02.095
5. Wadhwa L, Shekhar C, Saini J, Chetani M. Successful obstetric outcome in dextrocardia with situs inversus and moderate pulmonary hypertension-rare case. Journal of Clinical and Diagnostic Research: JCDR. 2016;10(10):QD08. Doi: 10.7860/JCDR/2016/20429.8749
6. Supriya G, Saritha S, Madan S. Situs inversus totalis–a case report. IOSR J Appl Phys. 2013 May;3(6):126.
7. Tabry IF, Calabrese J, Zammar H, AbouKasem K, Akeilan H, Gharbieh N, Zinati H, Noureddine W, Tayah M, Khalidy LA, Yaghi M. Case report: off-pump total myocardial revascularization for dextrocardia and situs inversus. InThe Heart Surgery Forum 2001 Sep 1 (Vol. 4, No. 3, pp. E251-E253).
8. Nawaz A, Matta H, Hamchou M, Jacobez A, Trad O, Salem AHA. Situs inversus abdominus in association with congenital duodenal obstruction: a report of two cases and review of the literature. Pediatric surgery international. 2005;21:589-92. Doi: 10.1007/s00383-005-1412-y
9. Agirbasli M, Hamid R, Jennings III HS, Tiller GE. Situs inversus with hypertrophic cardiomyopathy in identical twins. American journal of medical genetics. 2000;91(5):327-30. PMID: 10766992.

10. Vingerhoets G, Gerrits R, Verhelst H. Atypical brain asymmetry in human situs inversus: gut feeling or real evidence?. Symmetry. 2021;13(4):695. Doi: 10.3390/sym13040695
11. Sharma VK, Juneja A. A Rare Association of Complex Cyanotic Congenital Heart Disease with Situs Inversus Totalis Complicated by Brain Abscess in an Adolescent–A Case Report. Medical Journal of Dr. DY Patil University. 2023;16(4):645-7. Doi: 10.4103/mjdrdypu.mjdrdypu_986_22

Additional information

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Ethical Review Board Approval: The study conformed to the ethical review board requirements.
Informed Consent: Written informed consent was obtained from the patient for case report publication.
Conflicts of Interest: In compliance with the ICMJE uniform disclosure form, all authors declare the following:
Financial Relationships: All authors have declared that they have no financial relationships at present or within the previous three years with any organizations that might have an interest in the submitted work.
Other Relationships: All authors have declared that there are no other relationships or activities that could appear to have influenced the submitted work.

AUTHORS CONTRIBUTIONS

Sr.#	Author's Full Name	Intellectual Contribution to Paper in Terms of:
1.	Muhammad Naveed Majeed	1. Study design, literature review, methodology & paper writing.
2.	Muhammad Fateen Rashed	3. Data collection/analysis, results and calculations.
3.	Qasim Bashir	6. Editing and quality insurer.