

PIERRE ROBIN SEQUENCE ASSOCIATED WITH ARACHNOID CYST: A RARE NEUROLOGICAL MANIFESTATION

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Written informed consent was obtained from parents for publication of this case. Ethical approval was taken from the institutional review committee.

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Abstract

Pierre Robin Sequence (PRS) is a rare congenital condition defined by the triad of micrognathia, glossoptosis, and upper airway obstruction, commonly accompanied by cleft palate. While PRS most often occurs as an isolated anomaly, it may also be associated with other systemic malformations, including cardiac, renal, and neurological abnormalities. The coexistence of PRS with intracranial lesions such as arachnoid cysts is exceedingly uncommon and scarcely reported in the literature. We describe a case of a neonate who presented at birth with classical features of PRS, along with significant feeding difficulties and respiratory distress. Neuroimaging with computed tomography revealed a large left temporoparietal arachnoid cyst, highlighting an unusual neurological association. Early recognition of central nervous system involvement in infants with PRS is critical, as such comorbidities can substantially worsen clinical outcomes and increase the risk of morbidity and mortality. This case underscores the importance of maintaining a high index of suspicion for associated neurological anomalies in PRS and advocates for comprehensive evaluation beyond the craniofacial features alone. Prompt diagnosis allows timely intervention and supports optimal management strategies. Furthermore, this report emphasizes the need for a multidisciplinary approach involving neonatology, neurology, radiology, otolaryngology, and pediatric surgery to ensure coordinated care. Awareness of rare associations such as arachnoid cysts can aid clinicians in improving early detection and long-term prognosis in affected infants.

INTRODUCTION

Pierre Robin Sequence (PRS) is defined as a triad of micrognathia, glossoptosis, and airway obstruction, commonly accompanied by cleft palate. The condition may be isolated or syndromic, with reported associations including

genetic mutations, neurological malformations, and craniofacial syndromes. Neurological findings such as Chiari malformation, cerebellar hypoplasia, and agenesis of the corpus callosum have been reported but remain uncommon.

Arachnoid cysts in association with PRS are exceedingly rare. This report presents a newborn with classical PRS associated with a large arachnoid cyst.

Case Report:

A 22-day-old male infant presented with feeding difficulty since birth. He was born to a G5P1 mother via emergency LSCS due to fetal distress. Postnatally, he developed

cyanosis and respiratory distress requiring NICU admission for 7 days. Examination revealed micrognathia, glossoptosis, and a U-shaped cleft palate. Systemic examination was otherwise normal. CT brain showed a well-defined hypodense, loculated lesion in the left temporoparietal region measuring $5.1 \times 4.3 \times 7.1$ cm with density 1.3 HU, consistent with an arachnoid cyst.



Discussion:

PRS was first described in 1891 and later published in 1923. While cleft palate and airway problems are common, neurological abnormalities remain rare but clinically significant. Mortality in PRS with neurological involvement has been reported as high as 40% compared to 7% in isolated PRS.

Arachnoid cyst association is extremely rare, with minimal literature documenting this relationship. Infants with PRS often face feeding difficulties, respiratory compromise, and failure to thrive, increasing the burden on healthcare systems. Multidisciplinary management involving neonatologists, geneticists, neurosurgeons, and maxillofacial teams is essential.

Conclusion:

This case highlights a rare association of PRS with an arachnoid cyst. Neurological evaluation should be routinely considered in infants with PRS to avoid missed diagnoses and improve outcomes.

REFERENCES

- Abualshamat A, Al-Agha A. Pierre Robin Sequence in a Child With Ectopic Kidney, Polysyndactyly, And Short Stature: A Case Report. *Cureus*. 2019 Dec 27;11(12).
- Karempelis P, Hagen M, Morrell N, Roby BB. Associated syndromes in patients with Pierre Robin Sequence. *International Journal of Pediatric Otorhinolaryngology*. 2020 Apr 1;131:109842.
- de Buys Roessingh AS, El Ezzi O, Herzog G, Broome M. Facial Cleft and Pierre Robin Sequence. In *Neonatal Surgery: Contemporary Strategies from Fetal Life to the First Year of Age* 2019 Apr 14 (pp. 73-99). Cham: Springer International Publishing.
- Lee J, Hida K, Seki T, Kitamura J, Iwasaki Y. Pierre-Robin syndrome associated with Chiari type I malformation. *Child's nervous system*. 2003 Jun;19(5):380-3.
- Stevenson DA, Carey JC. A novel MCA-MR syndrome with PRS and cerebellar hypoplasia. *Am J Med Genet A*. 2007;143A:2221-6.
- Logjes RJ, Haasnoot M, Lemmers PM, Nicolaije MF, van den Boogaard MJ, Mink van der Molen AB, Breugem CC. Mortality in Robin sequence: identification of risk factors. *European journal of pediatrics*. 2018 May;177(5):781-9.
- Almasri M, Kishta W, Abduljabbar FH, Arlet V, Saran N, Oullet J. Ischiospinal Dysostosis in a Child with Pierre-Robin Syndrome. *Case Reports in Orthopedics*. 2017;2017(1):8263536.
- Varadarajan S, Balaji TM, Raj AT, Gupta AA, Patil S, Alhazmi TH, Alaqi HA, Al Omar NE, Almutaher SA, Jafer AA, Hedad IA. Genetic mutations associated with Pierre Robin syndrome/sequence: a systematic review. *Molecular syndromology*. 2021 Mar 18;12(2):69-86.

