Case report

Child withNon-Syndromic Lambdoid Craniosynostosis:A rare presentation

Abstract

Nonsyndromic lambdoid craniosynostosis is a significantly rare type of craniosynostosis where one of the lambdoid sutures fuses prematurely. This craniofacial anomaly is most often diagnosed before the age of 12 months and therefore early treatment is important to prevent neurological dysfunction, developmental delay, further facial anomalies, as well as disturbances with the optic nerve therefore affecting the eyes. This is managed by performing a posterior open cranial vault reconstruction surgery as well as a lambdoid occipital reconstruction. We report a rare case of non syndromic right lambdoid craniosynostosis in a 19 months old girl.

Categories: Neurosurgery, Pediatrics, Neurology

Keywords: Lambdoid craniosynostosis, non syndromic lambdoid craniosynostosis, craniofacial anomaly, lambdoid occipital reconstruction, posterior open cranial vault reconstruction

Introduction

Craniosynostosis occurs due to the premature fusion of one or multiple cranial sutures[1,4,9]. It can occur due to an association with other syndromes, classified as syndromic, or as an isolated condition known as nonsyndromic[2,4]. Types of craniosynostosis can be classified based on the affected suture which includes the sagittal, coronal, metopic, and lambdoid in order of frequency. These sutures allow the baby's brain to grow and expand as they develop. Therefore, early fusion of any of these sutures will present with an abnormal shaped head and in this case, positional plagiocephaly, which presents in lambdoid craniosynostosis. Failure of early surgical intervention and helmet placement will result in an increase in intracranial pressure leading to neurological problems, developmental delays, as well as eye, respiratory, and sensory dysfunctions [2,4,10]. Therefore, early surgical intervention before the age of 12 months is heavily advised. In this case of lambdoid craniosynostosis, the child was misdiagnosed at 6 months, during her regular checkup, with uncomplicated positional plagiocephaly which led to the continuous growth of the child's head in an abnormal

shape until the age of 19 months without any intervention. This rare case of non syndromic right lambdoid craniosynostosis is the least common type of craniosynostosis affecting around 1 in 150,000 births [1,9,10]. We present this rare case of a 19 months old girl with non syndromic right lambdoid craniosynostosis.

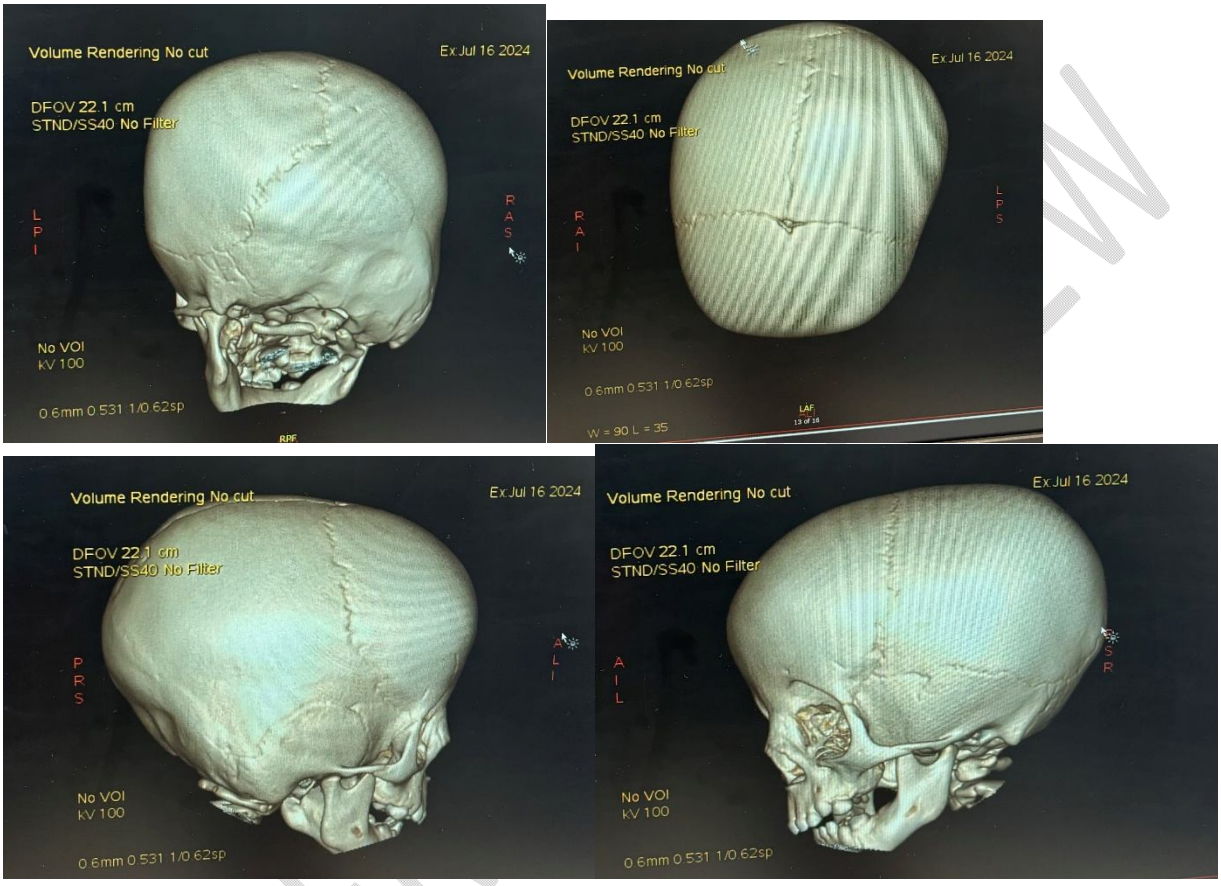
Case Presentation

An otherwise healthy 19 months old girl presented to the neurosurgery department for her delayed presentation of an abnormal head shape that became prominent the past month. She had no history of fever, vomiting, headaches, seizures, lethargy, trauma, or any past medical history or surgeries. She had no pertinent family history and she was born via normal delivery at 39 weeks. Her growth chart was plotted showing her height and weight to be in the 50th percentile along with her head circumference being 47 cm. She appeared to be alert, active, and well oriented and upon neurological examination she showed to have met her mental developments for speaking and walking and showed no developmental delays or neurological problems. Her gait examination was normal. She had no focal deficit and her systemic examination findings revealed to be unremarkable. She does not have any medical conditions, allergies, and is not on any medication. On examination she presented with a cranial deformity showing a flat forehead with supraorbital recession. She presented with left occipital bulging, bulging in the right mastoid area, and her right ear appeared to be inferiorly displaced. There was no cleft palate seen. She was referred to a pediatric ophthalmologist to further check for any symptoms of raised intracranial pressure which can add pressure to the optic nerve. Her eye examination showed good visual behavior and no refractive error outside of what is considered normal for her age. She showed healthy optic discs and no swelling. The pediatric neurosurgeon then recommended an x-ray of her skull using the technique of a helical scan of the brain without intravenous contrast with sagittal and coronal reconstructions. The 3D images revealed an abnormal right sided occipito-parietal flattening with contra-lateral bossing. It also clearly revealed the unilateral closure of the right lambdoid suture, whereas the other remaining sutures and fontanelles appeared normal. The ventricles, sulci, and basal cisterns all appeared normal and no signs of space occupying lesions were present. There was also mucoperiosteal thickening present in the right maxillary sinus and fluid in the right mastoid air cells.

She was diagnosed with right-sided lambdoid craniosynostosis consistent with posterior synostoticplagiocephaly. A plan for surgery was made which was concluded to be a posterior open cranial vault reconstruction and the risks and complications of the

procedure were thoroughly explained. Patient was operated but follow up was lost as the patient migrated to other country

Figure 1: a) X-ray of skull showing fusion of right lambdoid suture



Discussion

Craniosynostosis occurs due to the premature fusion of one or multiple cranial sutures.usually it is idiopathic but cases have been reported due to genetic causes; C S Fonteles , R H Finnell , et.al-reportes a similar case due to ALX4 mutation[1].

Surgical treatment of craniosynostosis is recommended at a very early stage and advised to be performed before the age of 12 months. The goal of early intervention is to reconstruct the cranial bone structures to allow the brain of the child to have the space to grow and expand as they develop.

Children with untreated craniosynostosis suffer from increased intracranial pressure, facial and cranial abnormalities, impairments of vision, hearing, developmental delays, and psychological disturbances [2,5,7]. Surgical intervention still remains to be the main treatment of craniosynostosis, however, there are still many complications with surgery that can occur such as infections, catastrophic bleeding, and death [3,4,6].

There has still not been any definitive cause found of non syndromiccraniosynostosis. There have been studies, however, showing that it can result from unknown genetic mutations, teratogenic exposure, chromosomal abnormalities, or environmental factors [7]. Syndromic craniosynostosis can also be caused due to associations with multiple syndromes such as Apert and Crouzon [8,10].

This case was thoroughly investigated and researched and all other causes of the abnormal head shape were ruled out. Although there is no prevention for craniosynostosis, early surgical treatment is very important to prevent further problems. This was a very rare and lucky case of right lambdoid craniosynostosis that was finally diagnosed at the age of 19 months as further delay would have resulted in irreversible problems.

Conclusion

Craniosynostosis is a major and complex craniofacial disorder that can cause many neurological dysfunctions if there is no early intervention. Therefore, children presenting with abnormal head shapes should not be ignored and should be thoroughly examined with X-rays and MRI's when necessary. Surgical intervention remains to be the main treatment, however studies are recommended to explore and further research the definitive cause.

Ethical Approval:

As per international standards or university standards written ethical approval has been collected and preserved by the author(s).

Consent

As per international standards, parental written consent has been collected and preserved by the author(s).

Disclaimer (Artificial intelligence)

Author(s) hereby declare that NO generative AI technologies such as Large Language Models (ChatGPT, COPILOT, etc.) and text-to-image generators have been used during the writing or editing of this manuscript.

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