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Adrenal fusion anomaly

Sunil Bylappa Kumar, Prashant Basavaraj Mahalingashetti, Ramaswamy Anikode Subramanian, Suneeta Padhy

CASE REPORT

A 35-year-old female, G2P1L1, underwent an antenatal scan at 36 weeks. Large thoraco-lumbar neural tube defect and Arnold-Chiari malformation type II were detected in the fetus. A female fetus weighing two kg was delivered following induction for termination of pregnancy. Autopsy confirmed Arnold-Chiari malformation, and meningomyelocele at thoraco-lumbar region. Histology showed features suggestive of amniotic fluid aspiration in non-aerated lungs. A transverse band of tissue across the midline was noted above the kidneys, posterior to aorta. No separate adrenals were identified in superior poles of kidney (Figure 1A, B). Histologic examination of transverse band of tissue showed a capsulated tissue with cells displayed in zonal pattern. A compact cellular layer formed outer layer, columns of paler cells in the middle and smaller cells in cords on inner side consistent with adrenal gland morphology (Figure 2). This confirmed the diagnosis of adrenal fusion anomaly.

Figure 1: (A) Abnormal midline band of tissue in the retroperitoneum (Gross picture - view from posterior aspect), (B) Closer view.

DISCUSSION

Adrenal gland agenesis, hypoplasia and accessory gland are common congenital anomalies of adrenal glands. While fusion anomalies of kidney are relatively common (1 in 300 pediatric autopsies), congenital fusion of adrenal glands is a rare anomaly with only 65 cases reported in literature [1, 2]. Most of them are detected at autopsy [1] and few as an incidental finding in imaging studies associated with other anomalies [1, 2]. These anomalies are functionally normal [2]. The fusion occurs across the midline, with resultant horseshoe or butterfly shaped gland. Fusion can be preaortic or postaortic [1, 2]. Normal histology of fused glands implies that the defect in embryogenesis occurs probably at 5–7th week. Similar case reports confirm the constant association with midline central nervous system defects such as meningomyelocele and Arnold-Chiari malformation. This confers the postulation that adrenal glands develop from single primordial gland rather than separate right and left glands. Horseshoe adrenal is less common component of asplenia

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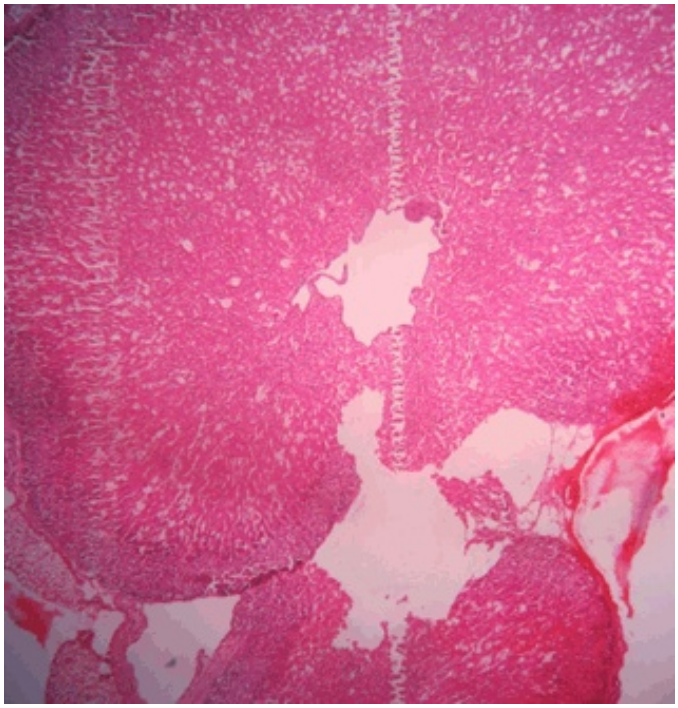

Figure 2: Cells arranged in three histological zones beneath the capsule (H&E, x100).

syndrome, a form of heterotaxy characterized by bilateral right-sidedness. Adrenal fusion may be considered as a differentiating feature between asplenia and polysplenia [3]. The management of such cases depends on type and severity of associated anomalies, their response to surgical intervention [2] and was detected antenatally which ends in termination as in our case. The occurrence of this anomaly in successive pregnancies is not mentioned in literature.

CONCLUSION

Congenital fusion of adrenal gland can be an incidental radiological finding. Awareness of this rare entity can avoid unnecessary interventions since they are histologically and functionally normal and no intervention is required but screening for potential assosciated anomalies such as central nervous system malformations, renal agenesis, asplenia, anomalies of the internal genitalia and complex cardiac anomalies are warranted.

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Author Contributions

Sunil Bylappa Kumar – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

Prashant Basavaraj Mahalingashetti – Conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Critical revision of the article, Final approval of the version to be published

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Guarantor

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Conflict of Interest

Authors declare no conflict of interest.

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