Diagnostic Imaging

Butterfly adrenal gland with maldevelopment of the mesonephric duct: A rare association in an adult patient

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ABSTRACT

Adrenal gland disorders can be asymptomatic and detected incidentally via imaging techniques such as ultrasound, computed tomography (CT), positron emission tomography, and magnetic resonance imaging. Fusion anomaly is a condition that can be attributed to errors in the developmental process and may be detected via these imaging modalities. We present a case of butterfly adrenal gland in a 61-year-old man with CT and magnetic resonance images. In our patient, this anomaly is also accompanied by unilateral renal agenesis and a diaphragmatic defect. Positron emission tomography-CT, contrast-enhanced CT, and magnetic resonance images are presented. To the best of our knowledge, this is the second case in which coexistence of unilateral renal agenesis and butterfly adrenal gland anomaly in an adult patient has been documented.

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Introduction

The adrenal glands were first described by Eustachius in 1563. Since then, important endocrine and physiological functions of adrenal glands and their pathologic conditions have been well documented.

Adrenal gland disorders can be of primary or secondary origin. Some of them are asymptomatic and detected incidentally via imaging techniques such as ultrasound (US), computed tomography (CT), positron emission tomography (PET), and magnetic resonance imaging (MRI) [1,2].

Fusion anomaly is a condition that can be attributed to errors in the developmental process and may be detected via these imaging modalities. The terms “horseshoe” and “butterfly” are used in medical terminology to signify the fusion of bilateral adrenal glands [3]. Compared with horseshoe kidney, horseshoe adrenal gland is an extremely rare anomaly associated with major cardiac or neural anomalies and is more frequently detected in newborn infants or at fetal necropsy [4–10].

We present a case of butterfly adrenal gland in a 61-year-old man with CT and MR images. In our patient, this anomaly is also accompanied by unilateral renal agenesis and a diaphragmatic defect. PET-CT, contrast-enhanced CT, and MR images are presented. To the best of our knowledge, this is the second case in which coexistence of unilateral renal agenesis and butterfly adrenal gland anomaly in an adult patient has been documented.
has been documented. Additionally, the literature was reviewed for prior reports of butterfly and horseshoe adrenal gland anomaly.

Case report

Our patient was a 61-year-old man with known unilateral renal agenesis. A renal mass was detected incidentally on routine US examination. For further evaluation, dynamic CT and MRI scans were obtained and revealed a solitary mass measuring 4 cm in the axial plane in the left kidney (Fig. 1), right renal agenesis, congenital absence of right seminal vesicule, diaphragmatic defect (Fig. 2), and fusion of bilateral adrenal glands (Fig. 3). The diaphragmatic defect was detected at the level of the aortic hiatus and was accompanied by hypertrophy of the diaphragmatic muscle adjacent to the defective area (Fig. 2).

Discussion

The adrenal glands are located adjacent to the diaphragma and the upper poles of the kidneys. The shape of the right gland resembles a pyramid. The left gland is semilunar in shape and larger than the right one [11]. They are composed of cortex and medulla. Fetal cortex derives from mesoderm; medulla

The adrenal glands were seen to fuse across the midline in front of the aorta. MR images demonstrated the same findings (Fig. 4). There was no pathologic fluorine-18-fluorodeoxyglucose uptake on PET-CT. The patient was asymptomatic, and there were no clinical or laboratory findings indicating adrenal dysfunction. Because the patient refused surgery, there is no pathologic diagnosis of the left renal mass.
originates from neural crest [12]. It is an important point that the adrenal glands develop separately from each other. Fusion mechanism is unknown but associated with different developmental pathologies. Adrenal fusion can occur in a pre- or post-aortic location [4] and may be detected via US, CT, and MRI.

Only 4 cases of horseshoe adrenals in asymptomatic adult patients are reported in the literature [3,13–15]. The first case of an adult patient with horseshoe adrenal glands was detected via CT imaging by Feldmann and colleagues in 2009 [13]. They also detected a posterior midline diaphragmatic anomaly in their patient. In 2013, Ditkofsky and colleagues presented a case of a female patient with similar findings; that patient was also asymptomatic, showed a posterior midline diaphragmatic hernia, a butterfly shaped ninth thoracic vertebra, and a unilaterally underdevelopment of the paraspinal musculature at the level of diaphragmatic defect [3].

In our patient, there is also a diaphragmatic defect, which is supporting the hypothesis of Ditkofsky et al. and Feldmann et al. [3,13]. These authors emphasized a similar embryologic precursor, anatomy, and temporal development of adrenal glands and diaphragma.

Another reported case in literature is that of a 22-year-old female who had a retroaortic located fusion of adrenal glands, congenital lumbosacral spina bifida with meningomyelocele, hydronephrosis of the right kidney, and a rudimentary uterus with imperforate hymen [14].

The fourth case showed a congenital absence of the right kidney, ureter, and seminal vesicle, as seen in our patient. The patient had also a left hydronephrosis secondary to ureteropelvic junction stricture. Apart from these findings, congenital fusion of the L4-L5 facets and a limbus vertebra at L4 were also detected in this patient [15].

Thus, our case is the second case reporting an association between butterfly adrenal gland and congenital absence of the right kidney, ureter, and seminal vesicle in adults.

**Conclusion**

We reported the second case of combined unilateral renal agenesis and horseshoe adrenal gland anomaly. Presumably associated with anatomic and developmental features of adrenal glands, congenital fusion is always seen with additional anomalies. Major abnormalities are often associated with horseshoe adrenal glands in fetuses or newborns. In our case, there was also a diaphragmatic defect, which emphasizes the theory that horseshoe adrenal gland anomaly may be caused by a failure of midline structures to separate. If horseshoe adrenal gland anomaly is detected on CT and MR images, radiologists should be aware of other concomitant developmental anomalies.

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**Fig. 4** – Congenital fusion of the adrenal glands. Axial T2 fat-saturated (A) and T2-weighted (B) magnetic resonance (MR) images demonstrate the “butterfly” shape of the adrenal glands.
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