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Abnormal abdominal and pelvic vasculature in a patient with Mayer-Rokitansky-Kuster-Hauser Syndrome.

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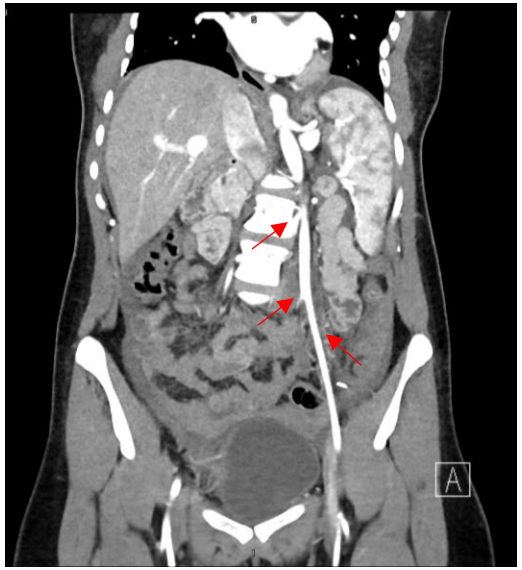
Background

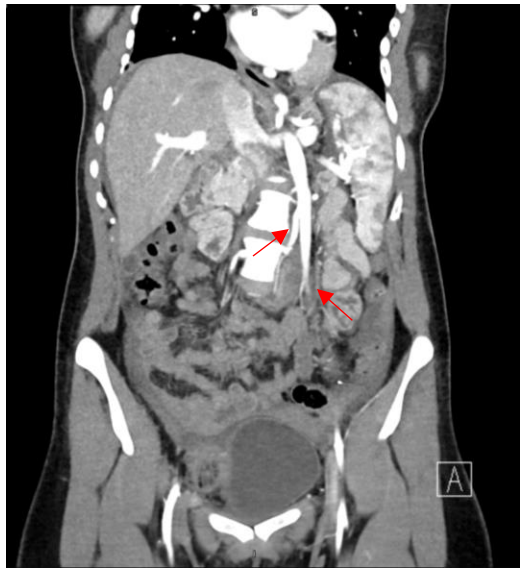
Mayer-Rokitansky-Kuster-Hauser (MRKH) syndrome, also known as Mullerian agenesis, is a congenital syndrome characterised by a failure of the development of the Mullerian duct, with a number of associated abnormalities affecting primarily (but not exclusively) the urogenital tract. Whilst MRKH is more commonly associated with renal, spinal and limb abnormalities, there have been much fewer cases of vasculature abnormalities described in literature.

Case

A 26 year old woman presented to hospital with severe pelvic pain following bilateral transabdominal oocyte retrieval for assisted reproduction earlier that day. Notably, the patient's past medical history included MRKH syndrome, with a congenital absence of the right kidney. The patient had previously undergone multiple procedures to create a neo-vagina, which had been complicated by fistulae and necessitated the formation of a colostomy. Given her medical background and unusual anatomy, a transabdominal approach to oocyte collection was used over the more conventional transvaginal approach. Whilst the procedure itself was uncomplicated besides the intraoperative finding of hypermobile ovaries, her pain in the hours following was concerning for a developing haematoma. A CT mesenteric angiogram demonstrated a right rectus sheath haematoma



Arterial phase CT mesenteric angiogram. Arrows indicate right internal iliac artery arising directly from aorta at level of L2, right external iliac artery arising from aorta at level of L3, and left common iliac artery.



Arterial phase CT mesenteric angiogram. Arrows indicate right internal iliac artery arising directly from aorta at level of L2, and bifurcation of aorta into right external iliac artery and left common iliac artery.

with active venous bleeding, as well as multiple vascular anomalies. The patient was referred for angiogram, which further characterised the vascular anomalies including a high aortic bifurcation, with the right internal and external iliac arteries originating separately from the aorta at the level of L2-3. During the angiogram no active bleeding was visualised, thus embolisation was not performed. Following the procedure the patient remained stable, and was discharged the next morning.

Discussion

The most common extragenital anomalies associated with MRKH syndrome are those of the renal tract, followed by vertebral and limb abnormalities. In some cases there may be hearing impairment, and rarely, cardiac anomalies. Though not commonly recognised as an association of this syndrome, abnormal vasculature has been described in a number of case studies, including one patient who had a similarly high bifurcating aorta.

This case highlights the importance of recognising the possibility of abnormal vasculature pre-operatively in patients with MRKH syndrome.

References

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