Case Report: Klippel-Trénaunay-Weber Syndrome

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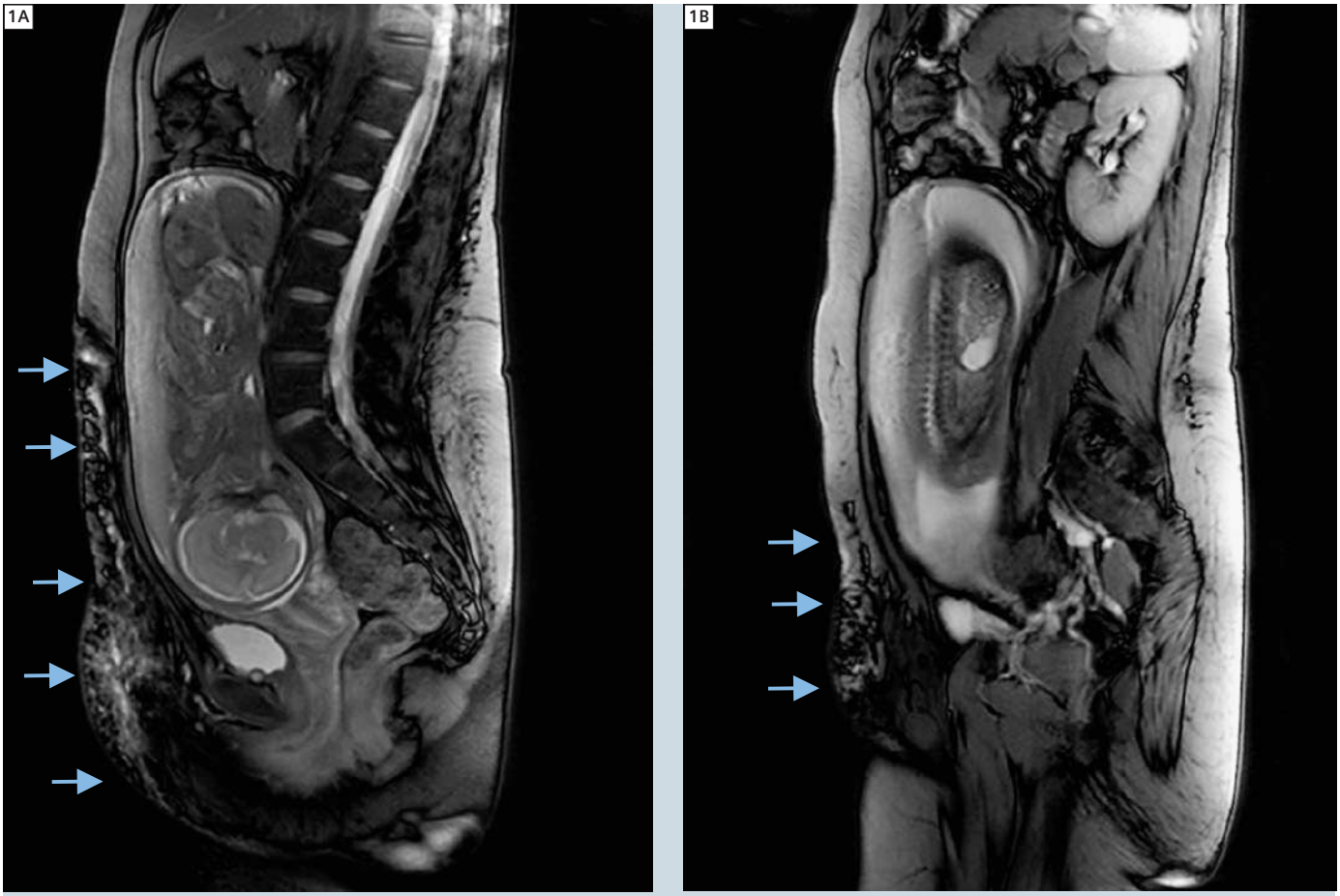
Patient history

We report on a 31-year-old female patient with known Klippel-Trénaunay-Weber syndrome. The patient was gravid and at 27 weeks gestation*. Referral to our institution was to confirm the presence of a venous malformation and aid obstetrical planning to determine if a Caesarean section was technically feasible.

The Klippel-Trénaunay-Weber syndrome is a rare congenital disease characterised

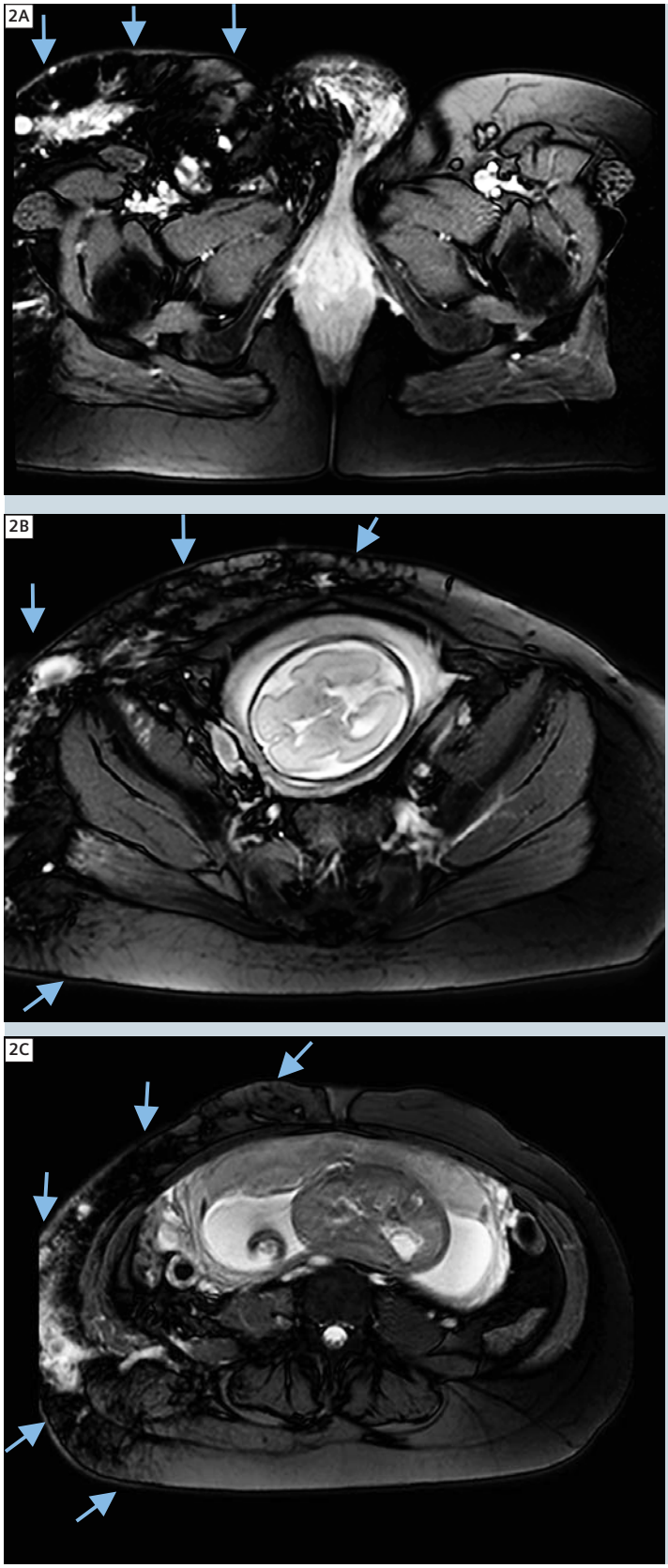


1 Composed sagittal TrueFISP images, consisting of two measurements. The extensive venous malformation is well delineated (arrows).

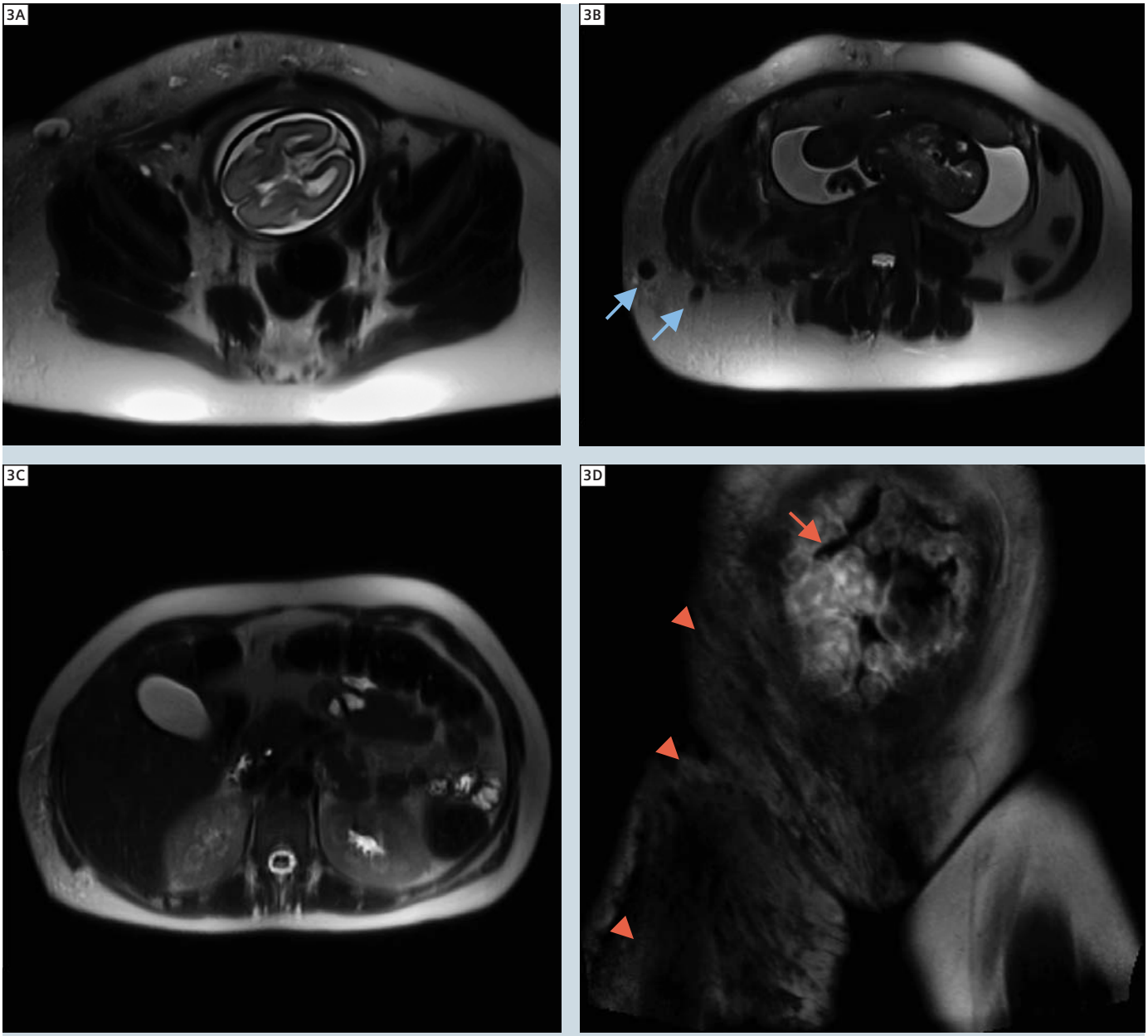
by a triad of capillary hemangioma (port-wine stain), large venous malformations as well as lymphangiomas. The haemangioma is of variable depth and can involve the deep sub-cutaneous tissues as well as involving adjacent visceral organs or bowel. It is also associated with focally limited gigantism (in very rare cases also dwarfism). The Klippel-Trénaunay-Weber syndrome is therefore also described as angiodysplasia with dominantly venous-cavernous type and hypertrophy of the affected extremity. Most cases are sporadic, although a few cases in the literature report an autosomal dominant pattern of inheritance. If arteriovenous malformations are present within the affected extremities, this special form is often described as Parkes Weber syndrome.

Sequence details

All images were acquired at 1.5T (MAGNETOM Avanto, 32-channel, SQ-Engine) with the usage of two Body Matrix coils and the integrated spine coil. The anatomical area covered with this MR exam reached from the upper lower extremities towards the upper abdomen. The examination was performed as a multistep MR exam. The scanner is equipped with the Tim Planning Suite. Because of the limited breath-hold capabilities of the patient, short and robust imaging sequence were mainly used (HASTE, TrueFISP). The protocol comprised: transversal (TR / TE 1380 / 76 ms, FOV 317 x 315 mm, matrix 220 x 256 Px, SL 7 mm), coronal (TR / TE 1375 / 76 ms, FOV 350 mm, matrix 243 x 256 Px, SL 7 mm) and sagittal (TR / TE 1380 / 76 ms, FOV 350 x 350 mm, matrix 243 x 256 Px, SL 7 mm) HASTE and TrueFISP (TR / TE 3.69 / 1.85 ms, FOV 350 x 350 mm, matrix 486 x 512 Px, SL 7 mm, respectively). For evaluation of potential large arterial feeder of the large vascular malformation, a dynamic MR sequence was used (3D FLASH in coronal orientation, 41 measurements, temporal resolution 6 seconds, TR / TE 2.13 / 0.76 ms, FOV 450 x



2 Transverse TrueFISP images. The involvement of the right thigh, pubis and abdominal wall by the vascular malformation is well visualised.

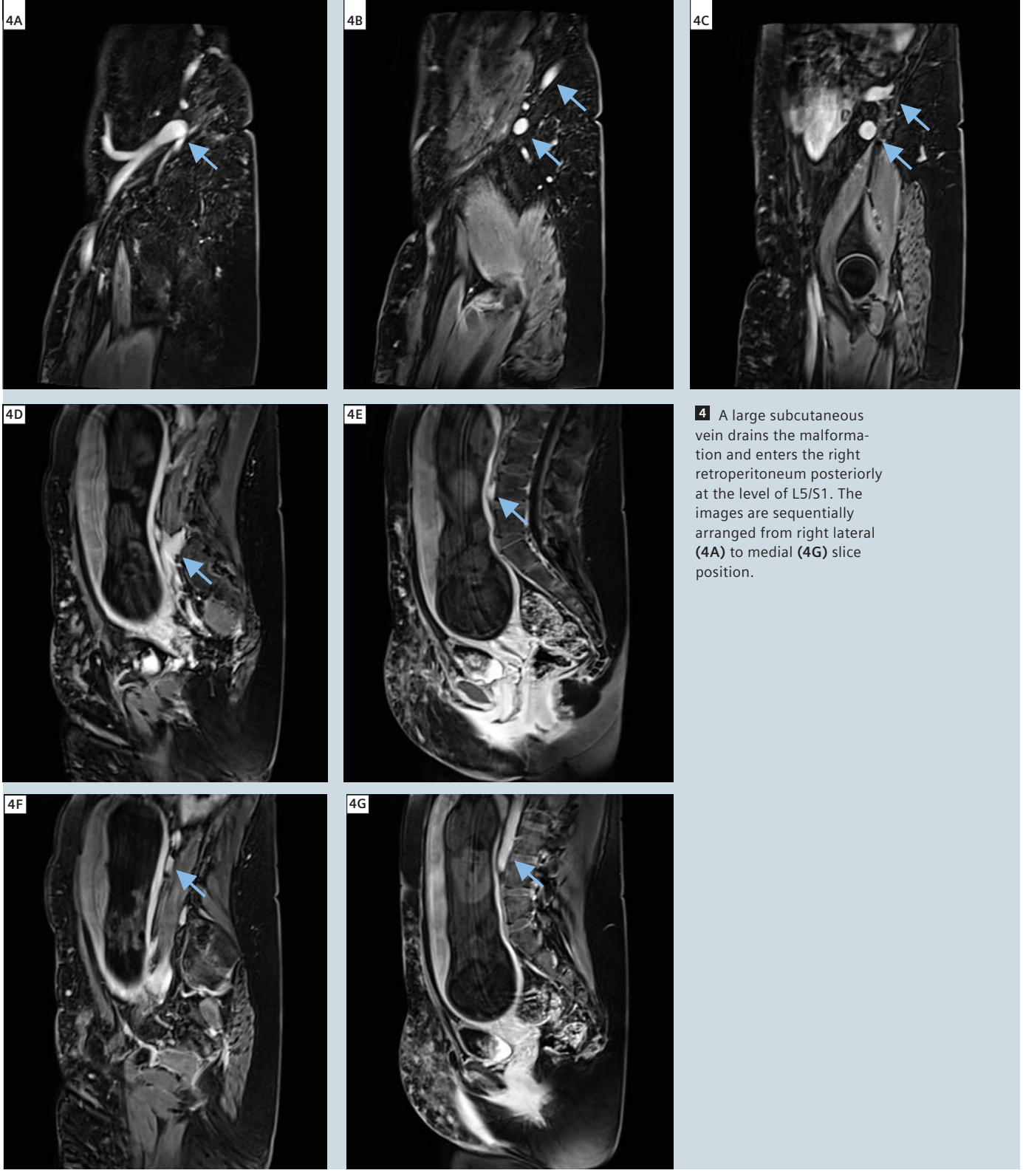


3 Corresponding transverse HASTE images **(3A–C)** show bright signal of the vascular malformation, indicating a low flow. However, two large vessels are shown with a clear flow-void (arrows, **3B**). Based on dynamic MRI, no arteriovenous malformation can be detected within the large vascular malformation (red arrowheads). Enhancement of the placenta is noted (red arrow).

450 mm, matrix 240 x 320 Px, SL 7 mm). Venous drainage was visualized by applying a 3D VIBE in sagittal orientation after contrast-media injection (TR / TE 5.41 / 1.72 ms, FOV 400 x 400 mm, matrix 240 x 320 Px, SL 5 mm).

Imaging findings

An extensive subcutaneous slow flow vascular malformation extending from the right thigh involving the right vulva, pubis, right lower abdomen and right flank was found. A large subcutaneous vein drained the malformation and entered the right retroperitoneum posteriorly at the level of L5/S1. An additional large subcutaneous vessel was identified in the right lower abdominal wall, which was favoured to comprise part of the venous drainage of the large malformation. No pelvic vascular malformations were seen. Also based on the dynamic MR scan, no evidence of an arterio-



4 A large subcutaneous vein drains the malformation and enters the right retroperitoneum posteriorly at the level of L5/S1. The images are sequentially arranged from right lateral **(4A)** to medial **(4G)** slice position.

venous shunt was found. The venous malformation crossed the midline to the contralateral abdominal wall; however, there were no venous malformations visible on the left side of the abdomen.

*The safety of imaging fetuses has not been established.

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