A 42-year-old man was admitted to Keiaido Hospital for sacral pain and dysesthesia in the segmental area S2 of the right lower extremity. He had been operated on for bilateral ectopia lentis. His uncle became blind by ectopia lentis and had undergone surgery for an aortic aneurysm. Multidetector-low CT (MDCT) demonstrated the dilated sinuses of Valsalva (Fig. 1). A diagnosis of Marfan syndrome was made. Further, lumbosacral magnetic resonance imaging (MRI) demonstrated a cystic lesion at the right S2 level (Fig. 2A and 2B). Communication with the subarachnoid space was revealed on computed tomography (CT)-myelography scan (Fig. 2C). A laminectomy of the sacrum was performed because S2 radiculopathy due to a sacral arachnoid cyst was suspected. A dark blue, cystic mass arose from the S3 root sheath and compressed the S2 root (Fig. 3A). The cyst was totally removed. Histopathological examination of the cyst wall revealed the features of a type I arachnoid cyst (Fig. 3B).

Marfan syndrome is an autosomal dominant disorder of connective tissue that affects the ocular, cardiovascular and skeletal systems. Meningeal abnormalities such as dural ectasia or anterior sacral meningocele are also seen in Marfan syndrome (1, 2). However, arachnoid cysts are a rare feature (3, 4). Arachnoid cysts are diverticula of the spinal meningeal sac or nerve root sheath and can be divided into three categories: extradural cysts without nerve root fibers (type I), extradural cysts with nerve root fibers including perineurial cysts (type II), and intradural cysts (type III) (5). Most spinal extradural cysts usually communicate with the arachnoid space by either a pedicle or an ostium, which may act as a check valve, creating an enlarged lesion (5). Further, hydrostatic pressure and transmitted pulsation of cerebrospinal fluid (CSF) promote expansion of the cysts (5). In patients with Marfan syndrome, weakened dura, due to deformed elastic tissue, also causes development of arachnoid cysts.
Figure 2. MRI and CT findings of sacral arachnoid cyst. The cyst at the S2 level (arrowhead) was isointense with CSF on sagittal T1-weighted image (A) and was hyperintense (arrowhead) to CSF in the lumbar region on a T2-weighted image (B). CT-myelography scan revealed contrast medium in the sacral arachnoid cyst (arrowhead) (C).

References

Figure 3. A) Operative finding. The arachnoid cyst (asterisk) arose from S3 nerve root sheath (arrow) and compressed the S2 root (arrowhead). B) Resected specimen of the cyst wall demonstrated a fibrocollagenous layer without nerve fibers (Masson stain, ×100).