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A case of Mullerian choristoma in extradural intra-extravertebral lipoma

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ABSTRACT

Objective: Mullerian choristoma associated with spinal dysraphism is one of the rare cases.

Case: We present a case of heterotopic fallopian tube tissue found in an extradural intra-extravertebral lipoma. The patient, a 19-year-old woman, complained of swelling and pain in her lower back. Magnetic resonance imaging revealed spina bifida occulta at L3, a tethered cord, and an L3 lipoma. A L3-L4 laminectomy was performed, and the lipoma was removed. Histological examination revealed the presence of ectopic fallopian tube tissue within the lipoma.

Keywords: Mullerian choristoma, spinal dysraphism.

INTRODUCTION

A choristoma is a benign, tumor -like growth of microscopically normal tissue that develops in an abnormal location (1). In this report, we present an unusual case of a 19-year-old woman who was diagnosed with Mullerian choristoma in association with spinal dysraphism, spina bifida occulta at L3, tethered cord, and L3 extradural intra-extravertebral lipoma. At the time of writing this text, there were fewer than 10 similar cases reported in the literature. (2)

CASE

A 19-year-old woman presented with a soft swelling in the lumbar region of her spine, which she had noticed since childhood. She also complained of low back pain, which worsened while walking. She did not have a history of any neurological deficits, and her menstrual history was normal. Upon physical and neurological examinations, there were no notable abnormalities, and routine laboratory tests were within normal ranges.

Further imaging tests were conducted, including a 1.5 Tesla MRI with IV contrast of the lumbo-sacral region of the spine. The MRI revealed spinal dysraphism, which is a congenital abnormality characterized by incomplete closure of the neural tube, at the level of L3 vertebra (**Figure 1**). Additionally, there was a tethered cord, which occurs when the spinal cord is abnormally attached to the surrounding tissue, and an extradural lipoma that was communicating with the subcutaneous fatty swelling. These findings were consistent with an extradural intra-extravertebral lipoma.

The patient underwent surgery, which involved a laminectomy at L3 and L4 and removal of the lipoma. The surgical specimen consisted of two pieces that measured 6.5 x 4 x 2 cm in total. Histological sections of the specimen showed skeletal muscles and mature adipose tissue with cyst-like structures that were lined with a single layer of columnar epithelium (**Figure 2**).

Immunohistochemical examinations were performed, and the results showed that the epithelial cells had a moderate positive nuclear reaction for PAX 8 and an intensive membranous reaction for CK7. The proliferative activity of the epithelial cells was low, and Ki67 labeled only 0.5-1% of the cells. The epithelial cells were negative for CK20. The overall histological picture and the results of the immunohistochemical examinations were consistent with the presence of ectopic fallopian tube tissue (i.e., ectopic tissue from the Mullerian compartment) within the soft tissues.

The patient recovered well after the surgery, and there were no postoperative complications. She was discharged from the hospital without pain.

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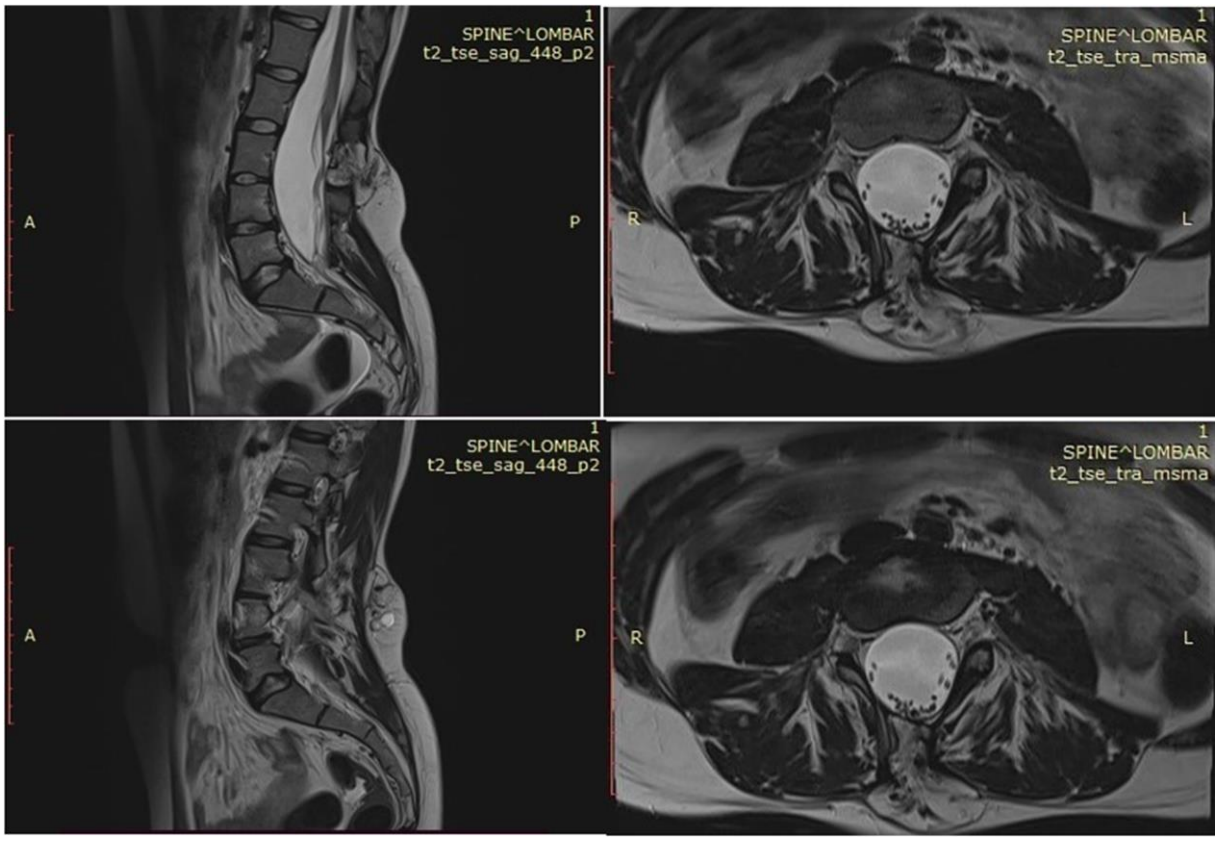

Figure 1: Figure 1 shows a 1.5 Tesla MRI with IV contrast of the lumbo-sacral region, revealing spinal dysraphism, spina bifida occulta at the level of the L3 vertebra, a tethered cord, and an extradural lipoma communicating with subcutaneous faty swelling.

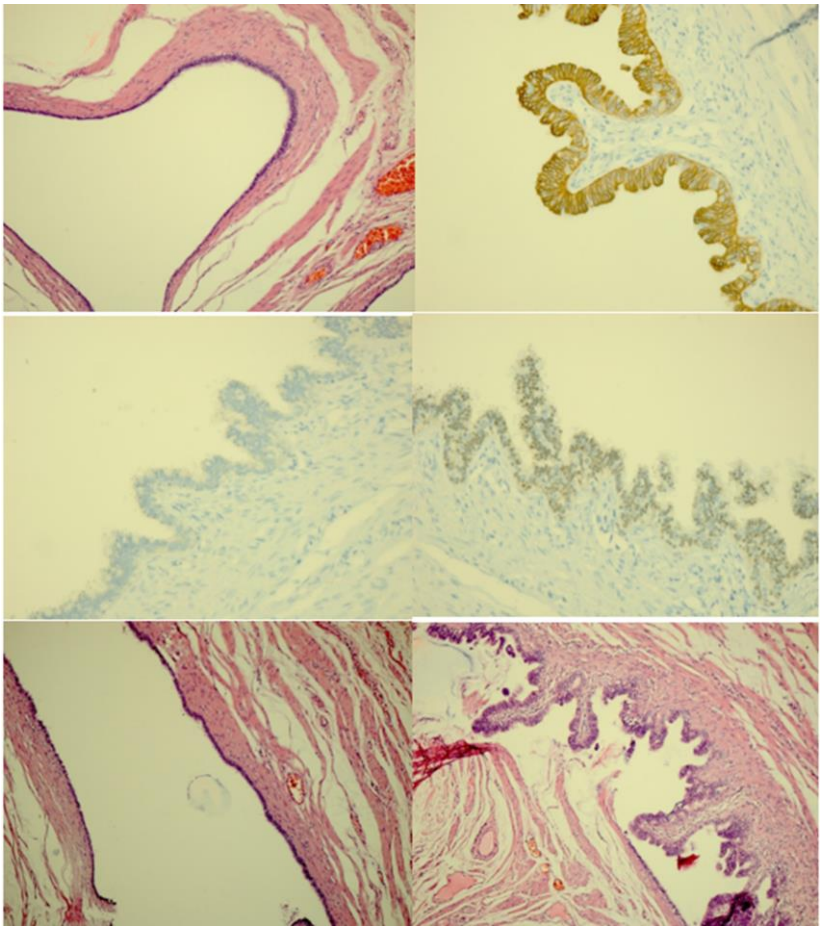

Figure 2: Histological and immunohistochemical examinations of the specimen showed the presence of ectopic fallopian tube tissue, which is a type of ectopic tissue from the Mullerian compartment, within the soft tissues.

DISCUSSION

The Mullerian ducts play a crucial role in the development of the urogenital system, giving rise to the uterus, uterine tubes, cervix, and the upper third of the vagina in females, under the influence of Anti-Mullerian Hormone (AMH). The development of the Mullerian ducts is tightly regulated by various signaling molecules and gene expression, including EMX2, HOXA13, PAX2, LIM1, and Wnt. These genes are also involved in the development of heterotopic sites, which can lead to the formation of Mullerian choristomas.

Spina bifida is a type of congenital malformation that occurs due to incomplete closure or formation of the embryonic neural tube, resulting in the splitting (bifid) of the spinal column (5). During primary neurulation, the ectoderm layer above the notochord undergoes proliferation to form the neural plate. The neural folds subsequently elevate from the lateral edges of the neural plate, ultimately approaching one another in the midline and fusing to create the neural tube.

During embryonic development, the process of neurulation involves the proliferation and elevation of the neural plate, which eventually fuses to form the neural tube. This process begins in the cervical region and progresses in both the cephalic and caudal directions. Below L-2, the spinal segments are formed by secondary neurulation. After neural tube closure, the epithelial ectoderm separates from the neural ectoderm through disjunction. The epithelial layers then fuse to form the skin covering the neural tube, while mesenchyme migrates between the neural tube and skin to create the meninges, neural arches of the vertebrae, and paraspinal muscles. In the third month of development, the spinal cord extends the entire length of the embryo. However, if there are any abnormalities in these developmental stages, a spinal dysraphism, or defect in neural tube closure, can occur (6).

While the exact cause of Mullerian choristomas is not fully understood, one hypothesis is that they result from misplacement of Mullerian duct tissue during embryonic development. Normally, the Mullerian ducts migrate towards the urogenital sinus to form the female reproductive tract. However, if some of the Mullerian duct tissue becomes misplaced and ends up in the caudal neural tube, it could explain the presence of Mullerian tissue in this location (7). This theory is supported by studies that have identified Mullerian-like tissue within spina bifida cysts and lipomas.

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Ethical approval: All procedures followed were in accordance with the ethical standards of the responsible committee on human experimentation (institutional and national) and/or with the Helsinki Declaration of 1964 and later versions. Informed consent or substitute for it was obtained from all patients for being included in the study. Written consent was obtained from each patient to use their hospital data.

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