A congenital “Pilonidal” Sinus at the tip of the coccyx - not with hair, but with atheroma

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CASE REPORT

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ABSTRACT

Introduction: Pilonidal Sinus Disease (PSD) is typically acquired, but there have been rare reports of congenital cases.

Methods: We describe the case of a young woman who experienced pressure and pain in the intergluteal region while sitting. The symptoms had been increasing over the years, with no signs of infection.

Results: MRI revealed a "pilonidal" sinus nest near the os coccyx, without a sinus extending to the skin. Surgery uncovered a deep-seated cyst near the tip of the coccyx, filled with atheromatous material but without hair. These findings are consistent with an epidermal fat cell that was translocated during embryonal development over the spine midline closure.

Conclusion: The rarity of this case is discussed, as the formation of congenital Pilonidal Sinus Disease is believed to be possible through the same mechanism.

Keywords: Embryology, skin closure, atheromatous tumor, pilonidal sinus disease, disease mechanism, congenital disease

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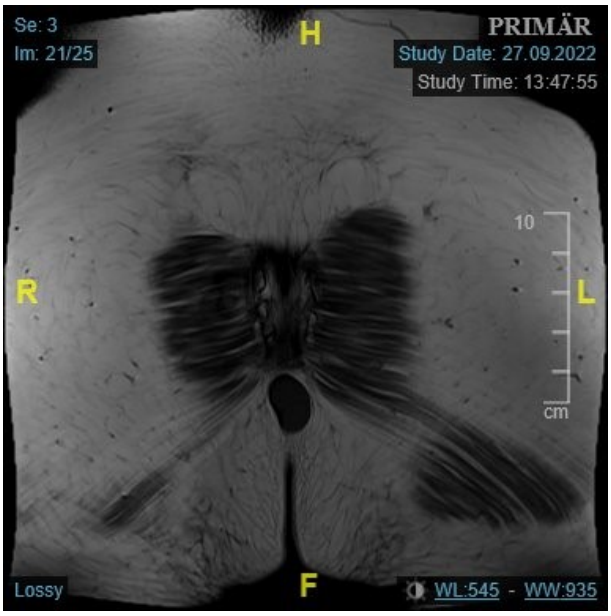
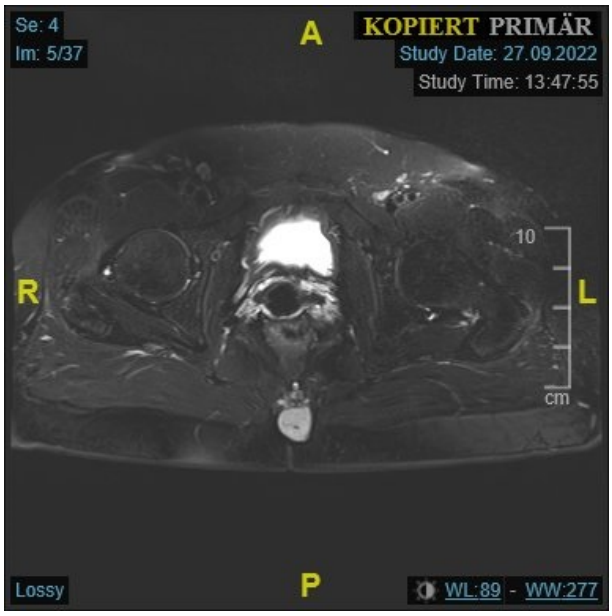
CASE REPORT

A 51-year-old female presented to our clinic with a persistent sensation of pressure in the coccygeal region, which had gradually progressed to pain while sitting. The patient reported no history of trauma, infection, or surgery in the glabellar and intergluteal region or lumbar spine. There was no family history of pilonidal sinus disease (PSD). On examination, a soft 2 x 2 cm mass was palpable in the depth of the intergluteal midline. Ultrasound and MRI revealed a cystic tumor with a fibrous capsule measuring 2.8 x 20 mm in close proximity to the coccyx, 1.8 cm from the skin surface, without involvement of the underlying bone (Figure 1 and 2). No midline sinus opening was observed on clinical examination or imaging.





FIGURES 1 and 2. MRI appearance

During surgery, a small, asymmetrical excision was used to approach the area, revealing a soft, elongated oval-shaped mass measuring 2 x 3 cm at the tip of the coccyx (Figure 3). The mass was not connected to the skin and was well-capsulated, containing white liquid fat without any hair. It was removed entirely, and the wound was left open to heal naturally. Histological analysis ruled out malignancy and described a fibrous capsule consistent with pilonidal sinus disease. Microbiologic examination confirmed that the contents were sterile. As can be seen in the Figure 3, the white liquid contained could be easily distinguished from the normal yellow fat surroundings, so a lipoma as an origin was judged to be highly unlikely. No sinus to the skin could be seen.

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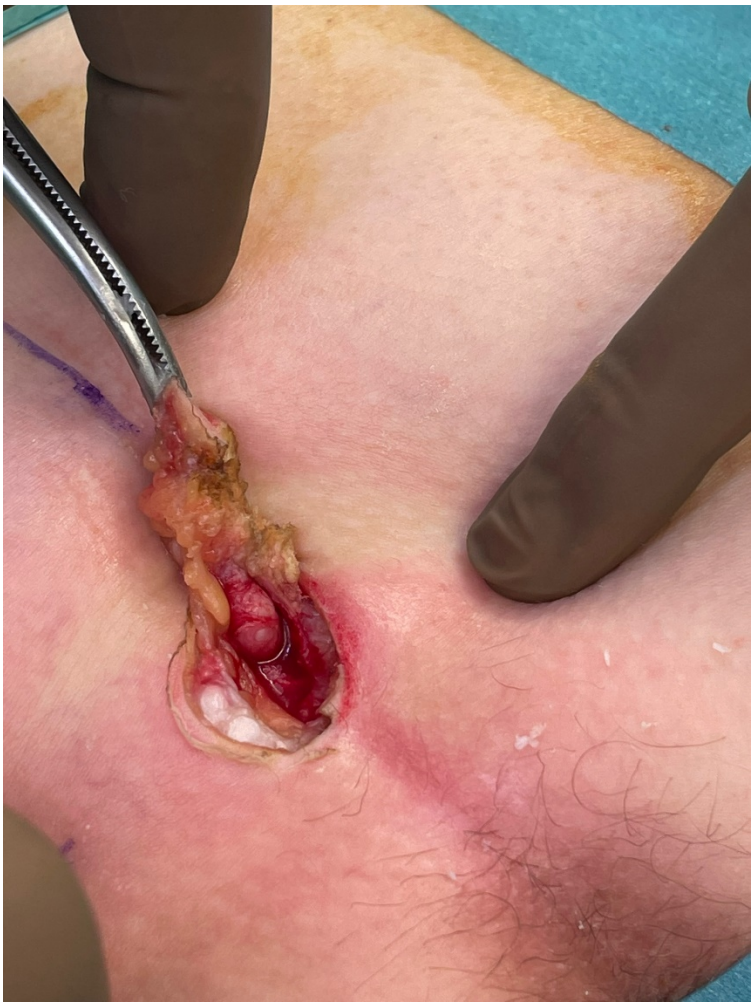

FIGURE 3. Intraoperative photograph with white liquid protruding from the saccular structure

For the first 110 years of its existence, pilonidal sinus disease (PSD) was believed to be a congenital condition. The midline location of the disease led embryologists to speculate that it was caused by a failure of ectoderm closure over the neural tube during weeks 3 and 5 of embryonic development. This theory was reinforced by the fact that spina bifida also occurred only in the midline. There have been reports of PSD occurring in identical twins (1-3) and families with a familial pilonidal trait (4-7). PSD communicating with the dura (8), as well as rare cases of PSD causing meningitis (9) and simultaneous cervical and lumbar PSD in one patient (8), have also been reported, which suggested a possible congenital origin.

In 1935, Fox proposed that either a persistent neurocutaneous opening or a faulty ectodermal invagination could lead to PSD (10). He used the presence of hair as evidence of its ectodermal origin, but there has been little proof of sweat glands or hair follicles in the depth. Tourneux and Herrmann postulated a coccygeal remnant being responsible for tumors and pilonidal disease around the os sacrum (11), but this theory could not be supported by histology (12).

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The theory of "jeep's disease" proposed by Buie in 1944 was a turning point for the acceptance of the acquired theory of PSD (13), even though mechanical warfare was not the reason (14) for the more than 78,000 PSD cases in the US Army between 1941 and 1944 (15). Calvet and Gontuier proposed that only 10% of all cases of PSD are of congenital origin (15), and Hodges believed that hair penetrating sacral skin leads to a subcutaneous foreign body reaction that ultimately forms the pilonidal sinus, but only if a congenital sacral dimple, trapping hair, is present (16).

Nowadays the acquired theory is not questioned any more, as sharp hair fragments falling down from the occiput (17) have been found to inject themselves into the upper third of the intergluteal fold, as clinical (18, 19), forensic biology (20) and electron microscopy studies (21, 22) have shown.

Nevertheless, congenital cystic disease did occur, but was exquisitely rare. This could be distinguished between cystic disease with or without connection to the neural tube, which could lead to meningitis. Therefore, a single hair follicle with a fat-producing cell may have been transferred to the tip of the coccyx and slowly enlarged over the decades into an oval-shaped tumor without infection or malignancy. The absence of hair in the specimen supports this theory.

Here we propose a single hair follicle with fat producing cell has been transferred onto the tip of the coccyx and has enlarged slowly over the decades into an oval shaped tumour without infection or malignancy. There was no hair in the specimen.

In the MRI-scan that has made the diagnosis before, there is unfortunately no possibility to measure density (as in CT). So, the cystic structure was taken as PSD at a typical location, with a tract not seen. In ultrasound, an inhomogeneous echo was seen, possibly related to the solid fat chips inside a liquid emulsion as shown in Figure 3.

CONCLUSION

Not every tumour overlying the sacral bone is a pilonidal sinus. If there is no tract present, and no infection (or swelling) present, it might be a case that a surgeon sees once in a lifetime. If there is a longstanding pilonidal sinus disease with tract and ulcer, a pilonidal sinus carcinoma may be present.

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MULTIMEDIA

N/A

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ETHICS

This study was conducted in accordance with the principles outlined in the Declaration of Helsinki, the analyses conducted in this study did not involve any interventions that could potentially cause harm to human participants. Written approval was obtained from the patients.

CONFLICT OF INTEREST

All authors declare that they have no conflicts of interest, and there are no relevant or minor financial relationships between relatives or next of kin and external companies.

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