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Case Report

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Glomus coccygeum in pilonidal sinus surgical specimens: report of two rare cases with special reference to SOX10 expression

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Abstract

We report two new cases of glomus coccygeum in pilonidal sinus excision specimens. The positive expression of glomus coccygeum cells for SOX10 is used for the first time. SOX10 is a useful immunohistochemical marker for identifying this microanatomical structure, confirming the diagnosis and may help the differential diagnosis. The glomus coccygeum cells are probably neural crest-derived from multipotent Schwann cell precursors.

Keywords: glomus coccygeum; pilonidal sinus; SOX10; vestigial structure; sacral area

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Introduction

Glomus bodies (including coccygeal glomus) consist of modified smooth muscle cells arranged in layers around small vascular channels.[1] They are a modified arteriovenous anastomosis in which innervation is similar to the canals of Sucquet-Hoyer of the distal phalanges of the toe.[2] When found in distal extremities, they generally do not represent a diagnostic problem.

The glomus coccygeum (GC) is a vestigial structure in the deep layers of the skin that is removed from the lumbosacral region. Its function in this area is still unknown.[3]

The GC may be an incidental finding in the tissue surrounding a pilonidal sinus cyst in surgical specimens.[4] The frequency of this topographic combination is low with only 11 cases (including our two cases) reported in the English literature.[3–5] It may represent a diagnostic challenge to the surgical pathologist.[2,5,6]

Immunohistochemically, the glomus cells express smooth muscle actin (SMA), vimentin, neuron-specific enolase (NSE) but do not express desmin, S100 protein or endothelial cell markers: CD31 and FVIIIR: Ag. Both S100-positive (nerve sheath) cells and CD31/FVIIIR: Ag-positive endothelial cells are typically present within the glomus body.[7]

The SOX10 transcription factor is known to be important in the development and maintenance of the peripheral nervous system.[8,9] The SOX10 nuclear protein is widely expressed in glial cells, melanocytes, Schwann cells, and myoepithelial cells.[10] To our knowledge, SOX10 has not been reported in GC in the medical literature so far.

Case Report

Case 1 was a 20-year-old man presented with symptoms of a pilonidal sinus cyst. Grossly, the local excision surgical specimen was 6×1×3 cm in size and contained a cyst with diameter of 1.8 cm.

Case 2 was a 45-year-old woman presented with an infected cutaneous lesion for 6 months with a recurrent abscess. Grossly, the cutaneous fragment was 2.5×1.5×1

cm in size, the surface of which was raised by a 1.5 cm cyst, with a creamy content.

Histological examination of both lesions showed the presence of a pilonidal sinus cyst. In both specimens, when a deep cut section of the fragments was done, glomus structures measuring 0.3 cm (case 1) and 0.5 cm (case 2) along the long axis were found. The lesions were recognized as sharply circumscribed complex structures composed of clusters and nests of small to medium-sized epithelioid cells associated with small vascular channels and with small nerve bundles (**Figure 1a**).

Immunohistochemistry of both lesions showed that glomus cells expressed SMA (**Figure 1b**), NSE and vimentin and were negative for desmin, cytokeratin, CD31 and CD34 (data not shown) and S100 protein (**Figure 2a**). Proliferative activity was low. Besides, in both cases, SOX10 - staining was presented in cellular nuclei of most of glomus cells (**Figure 2b**). The H-score was used for the interpretation of the SOX10 expression.[8] The median H-score for SOX10 in both cases of GC was 50 (40–60) (a marker was considered positive when its H-score was ≥10).[8]

Discussion

The GC is a non-pathologic structure that exhibits significant variations in size and proportion ofits constitutive elements. It is presented in every completely excised specimen of the coccyx.[1] This structure is composed of modified smooth muscle cells (glomus cells) arranged in concentric layers around blood vessels.[1,2] Immunohistochemical expression of SMA and NSE in glomus cells may be beneficial for accurate identification of a diagnosis. The results we have obtained for PS100- /SOX10+ immunophenotype of GC-cells support the fact that they are neural crest-derived, similar to glomus cells in the carotid body.[3] The very close association of these cells with Schwann cells probably supports their origin from multipotent Schwann cell precursors.[3,4]

Our observation contains a practical aspect in the differential diagnosis of normal or hyperplastic GC versus glomus tumor and paraganglioma. In the latter cases, SOX10 is not expressed or expressed only in the sustentacular cells, but not in the tumor cells. Apart from that, they originate from the neural crest, the positive spectrum of SOX10 in GC can probably be explained by the available myoepithelial component in these structures, which is also SOX10 positive. The lack of expression of SOX10 in the glomus tumor may be due to a P46S mutation which is not available in normal or hyperplastic GC.[11]

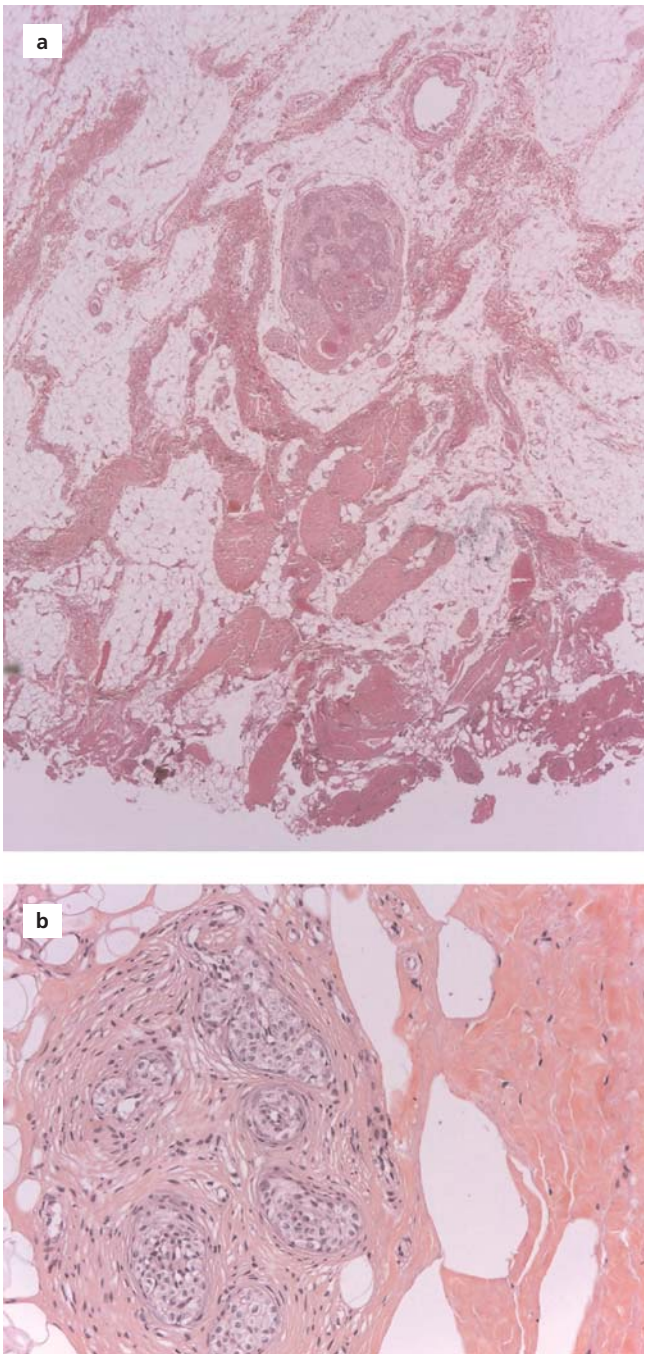

Figure 1. Sections stained with hematoxylin-eosin-saffron (HES) showing a sharply circumscribed complex structure (glomus body) situated deep in the connective tissue. (**a**) Case 1 with clusters and nests of small to medium-sized epitheloid cells associated with a small vascular channel, ×25 magnification; (**b**) Case 2, ×200 magnification.

In conclusion, we describe two new cases of the GC in pilonidal sinus excision specimens. SOX10 is a helpful marker for identifying this microanatomical structure and confirming the diagnosis. Adding our two cases to the lit-

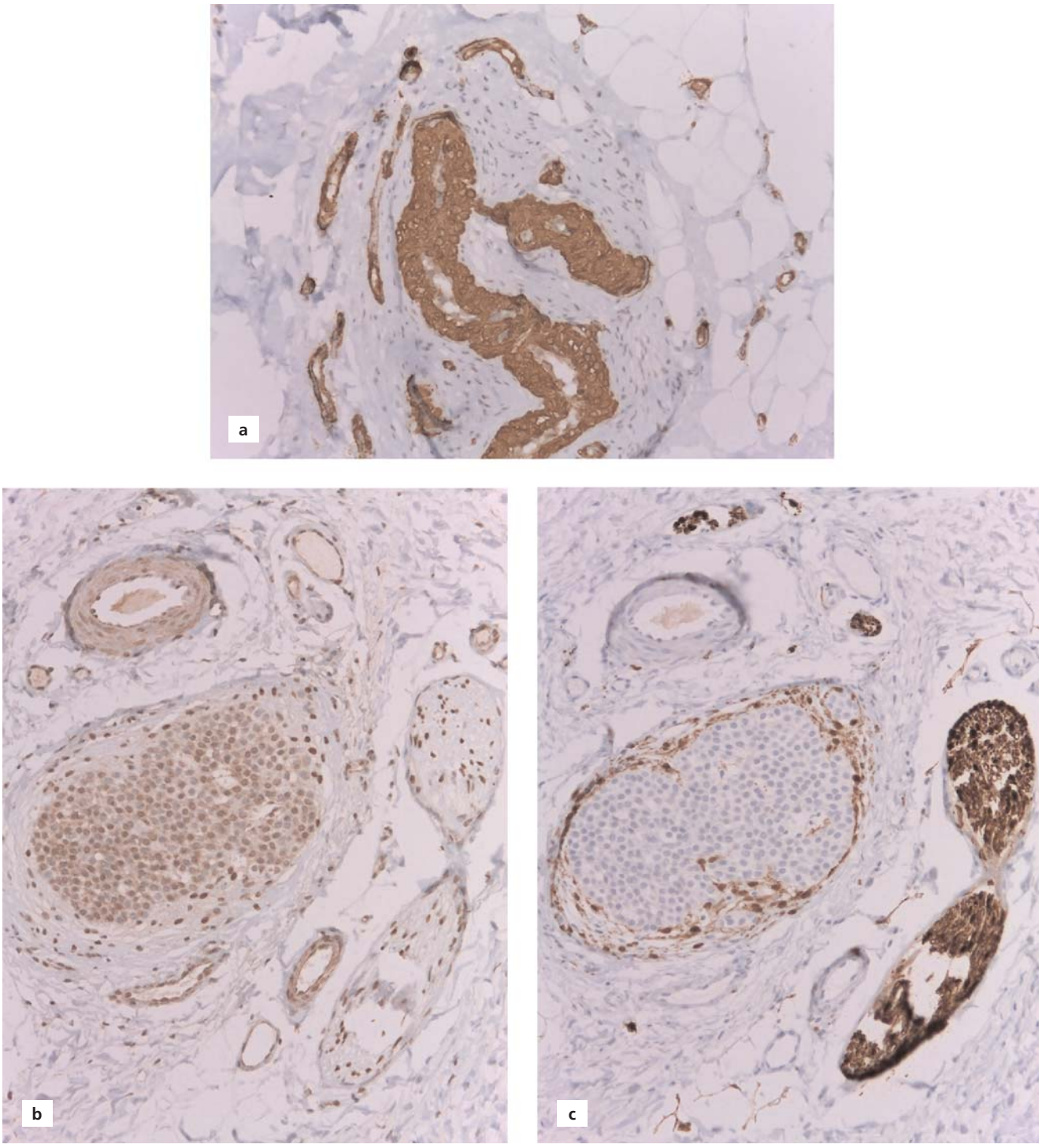

Figure 2. Immunohistochemical staining of glomus cells for smooth muscle actin. (**a**) Case 1, x200 magnification; (**b**) hyperplastic glomus cells lack expression of S100 protein; note S100 protein accentuated peripheral Schwann cells (Case 2), ×100 magnification; (**c**) glomus cells show variable intensity of SOX10 intranuclear staining (Case 2), ×100 magnification.

erature provides novel clinicopathological data, useful for precise diagnosis and avoids confusion with glomus tumors in the sacral area. To the best of our knowledge, the positive expression of GC- cells for SOX10 is used for the first time as a reliable immunohistochemical marker that may aid the diagnosis and the differential diagnosis.

Conflict of Interest

The authors declare no conflict of interest.

Author Contributions

SP: evaluation of the cases, writing the manuscript; IM: design of the study; MKI: writing the manuscript; DD: design of the study, supervision, critical revision of the manuscript.

Ethics Approval

The study was performed following the aid of the ethical standards down in the 1964 Declaration of Helsinki and its later amendments. Written and signed consent was obtained from the patients.

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