Colonic mucosal Schwann cell hamartoma with tactile corpuscle-like bodies: a case report

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A rare case of colonic mucosal Schwann cell hamartoma with tactile corpuscle-like bodies is described. It was a 4 mm polyp removed by colonoscopy in a 67-yr-old female patient without personal/family history of inherited syndrome. The lesion contained vague fascicles of bland Schwann cells and several tactile corpuscle-like bodies. Immunohistochemically, it expressed strongly S100 protein and SOX10, and it was negative for CD34, EMA, alpha-smooth muscle actin, CD99, CD56, chromogranin A, synaptophysin, c-kit, DOG1, and calretinin. This case supports an opinion that mucosal Schwann cell hamartoma and mucosal tactile corpuscle-like bodies of the gastrointestinal tract share a common histogenesis and that they represent a morphologic spectrum of a single entity.
Keywords: colon, mucosal Schwann cell hamartoma, tactile corpuscle-like bodies, immunohistochemistry

Hamartóm zo Schwannových buniek v sliznici hrubého čreva: popis prípadu
Popísaný je prípad hamartómu zo Schwannových buniek hrubého čreva, s tvorbou meissneroidných teliesok. Jednalo sa o 67-ročnú ženu, ktorá nemala klinické známky ani rodinnú anamnézu svedčiace pre dedičný syndróm s neuronálnymi proliferáciam (neurofibromatóza, MEN, adenomatózna polypóza). Pri kolonoskopii bol odstránený 4 mm-ový polyp sigmy. Histologicky obsahoval vretenobunkovú štruktúru Schwannových buniek, s málo početnými meissneroidnými telieskami. Imunohistochemicky bola lézia silne pozitívna na S100 protein a SOX10. Negatívne boli CD34, EMA, alfa-hladkosvalový aktín, desmín, CD99, CD56, chromogranín A, synaptofyzín, c-kit, DOG1 a kalretinín. Prípad podporuje v nedávnej literatúre prezentovaný názor, že mukózny hamartóm zo Schwannových buniek a meissneroidné telieska sliznice GIT-u majú spoločnú histogenézu a reprezentujú morfologické spektrum jedinej lézie.
Kľúčové slová: hrubé črevo, mukózny hamartóm zo Schwannových buniek, meissneroidné telieska, imunohistochémia

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Introduction

In 2009, Gibson and Hornick described a novel and rare colonic Schwann cell lesion that differs from already well-known neural lesions of the gastrointestinal tract, such as neurofibroma, schwannoma, ganglioneuroma, perineurioma and mucosal neuroma[(1)]. They termed it mucosal Schwann cell hamartoma (SCH). Subsequently, additional cases were published[(2-4)]. In the same year, Myisidis et al. described in gastric mucosa another neurogenic lesion which consists of tactile-like corpuscles[(5)]. A few similar cases found in colonic mucosa and in other gastrointestinal locations were described afterwards[(6-10)]. Quite recently, Cereilo Munoz et al. added further 9 cases and they labeled the lesion „tactile corpuscle like bodies of the mucosa"[(11)]. In this study, the authors shortly mention a possibility that tactile corpuscle like bodies could represent a variant of SCH. This is supported by one published case of SCH by Ferro de Beca et al. who described „tactoid body features" in the lesion[(12)]. We would like to demonstrate an additional SCH that contains tactile corpuscle like bodies. This observation supports further the opinion that SCH and tactile corpuscle like bodies are closely interrelated from histogenetic point of view and that both lesions may represent a morphologic spectrum of a single entity.

Case report

A 4-mm-sized polyp of the sigmoid colon was found in a colonoscopy of a 67-year-old female who has no significant family history of other neuronal lesions or inherited syndromes. The reason for colonoscopy was positive fecal occult blood test and anemia.

Histologically, the polyp showed a poorly circumscribed proliferation of bland appearing spindle cells in the lamina propria, separating the crypt architecture ***(Figure 1)***. The cells formed small plexiform nests and short ill-defined fascicles. Some of the nests showed concentric and lamellated arrangement of the cells, resembling closely that of Wagner-Meissner tactile body ***(Figure 1c)***. Nuclear atypia, pleomorphism, or mitoses were not observed. A few colonic crypt of the lesion were hyperplastic, with serrated morphology. On immunohistochemical staining, the spindle cells displayed a strong and diffuse positivity for S-100 protein

Figure 1. *Histological features of Schwann cell hamartoma with tactile corpuscle-like bodies.* ***(a, b)*** *Bland appearing Schwann cell proliferation in the mucosa forms ill-defined cell nests and fascicles.* ***(c)*** *At high power, some of the cell nests showed resemblance to Wagner-Meissner body.*

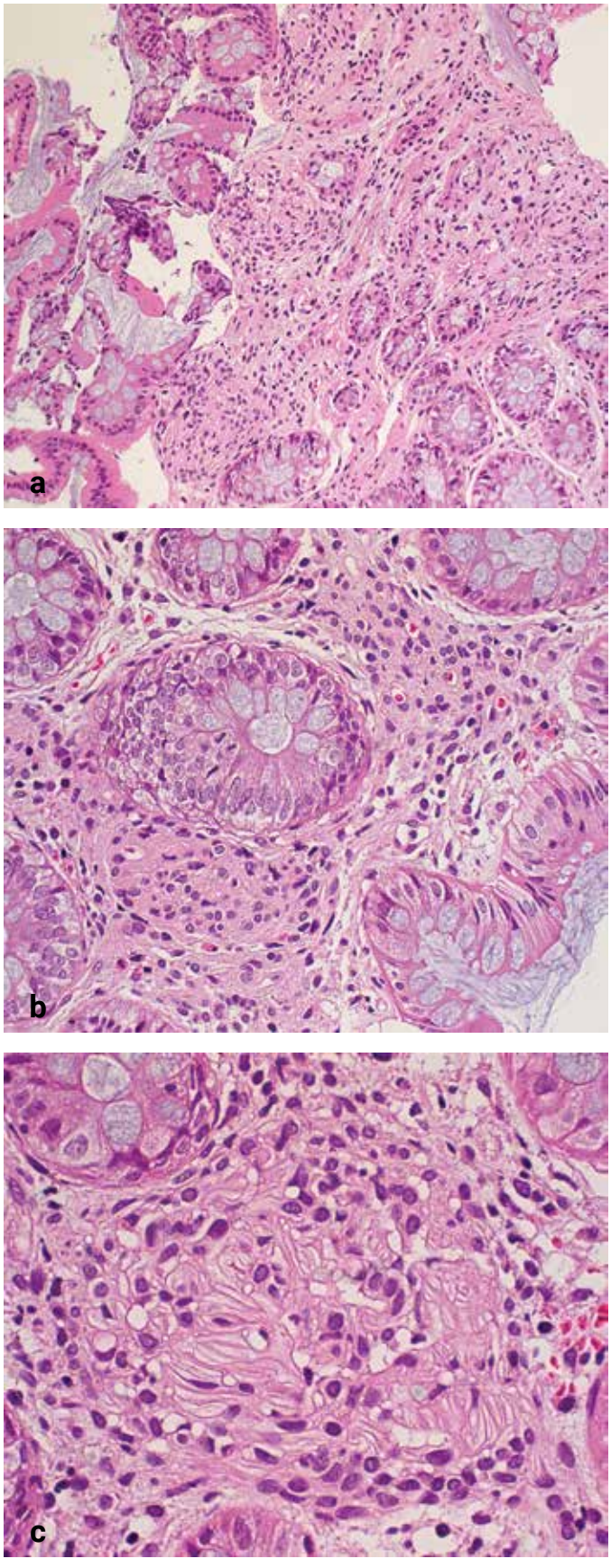
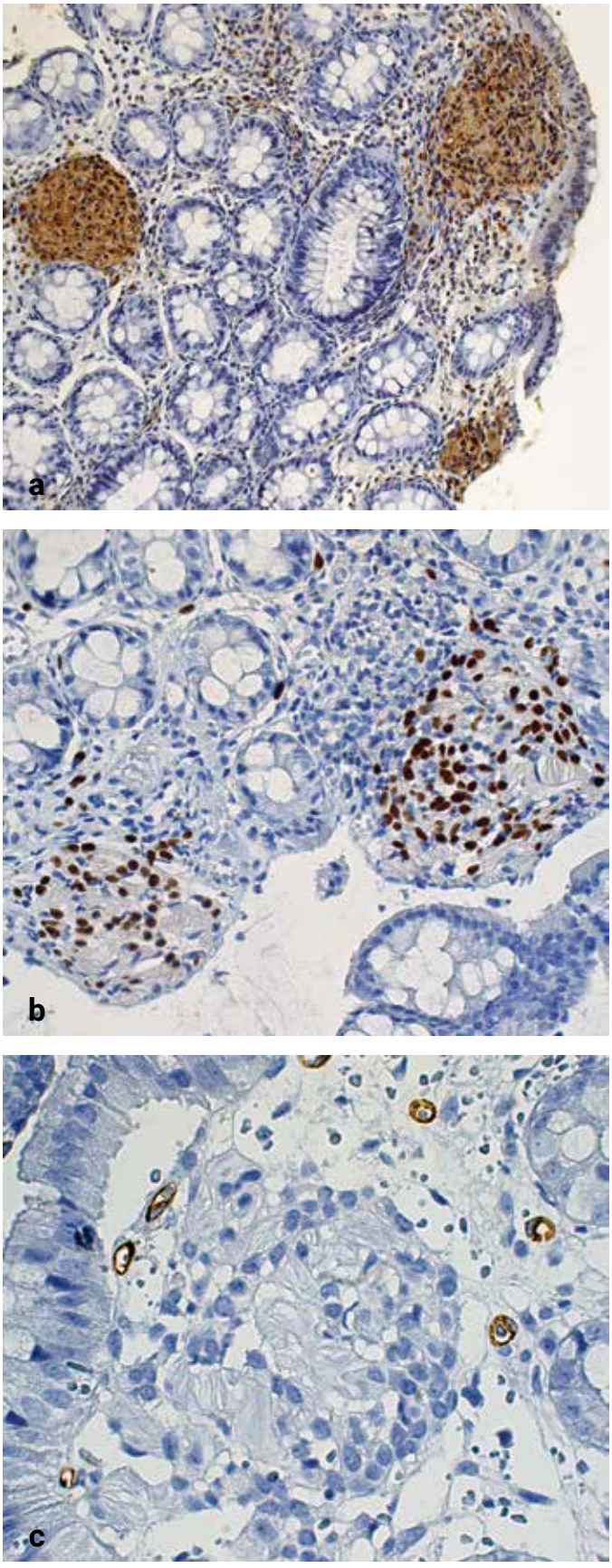


Figure 2. *Immunohistochemical features of Schwann cell hamartoma with tactile corpuscle-like bodies.* ***(a, b)*** *Positivity for S100 protein and SOX10 are shown, respectively.* ***(c)*** *CD34 is limited to the vessels and it is negative in the cells of the lesion.*



(polyclonal, DAKO) and SOX10 (clone EP268, Cell Marque) ***(Figure 2)***. They were negative for following antibodies: CD34 (QBend/10, prediluted, Biogenex) ***(Figure 2c)***, EMA (E29, prediluted, Biogenex), synaptophysin (Snp88, prediluted, Biogenex), alpha-smooth muscle actin (1A4, DAKO), CD99 (O13, Ventana), CD56 (123C3, DAKO), chromogranin A (DAK-A3, DAKO), c-kit (polyclonal, DAKO), DOG1 (SP31, Cell Marque), and calretinin (DAK-Calret1, DAKO).

Discussion

In our case, the tumor was composed of Schwann cell proliferation with features typical of SCH[1], i.e., non-circumscribed mucosal lesion composed of vague ill-defined fascicles of bland S100 protein positive spindle cells. In addition, some plexiform foci of the tumor formed lamellated structure with morphology of tactile corpuscle like bodies, as described by Myisidis and Cereilo Munoz at al.[5,11]. Our case is quite similar to the case of SCH with tactoid body features described by Ferra et al.[12]. Both cases indicate that SCH and tactile corpuscle like bodies may represent a single entity and that tactile corpuscle like bodies of the mucosa can be regarded as a special form of SCH. By reading of previous reports of SCH, we have realized that some of these studies show in SCH structures similar to the tactile corpuscle-like corpuscles, and therefore the cases could be considered to be SCHs with tactile corpuscle like bodies. Ortiz et al[2] in their figures 1 and 2, and Han et al[3] in their figure 1C depict round Schwann cell nests with lammellated cell arrangement which is quite similar to that of Wagner-Meissner corpuscle. In addition to this morphologic overlap between SCH and tactile corpuscle like bodies, it is apparent that both lesions overlap also from clinical point of view. They both appear to be reactive non-neoplastic, and all published cases lacked an association with known syndromes with gastrointestinal nerve sheath cell proliferations, such as type 1 neurofibromatosis and multiple endocrine neoplasia, type 2 B[1]. For this reason, it is important for pathologist to be familiar with these lesions. The differential diagnosis of our case included gastrointestinal stromal tumor[13], neurofibroma[14,15], mucosal neuroma[16], ganglioneuroma[17], mucosal schwannoma[18,19] and perineurioma[20,21]. Gastrointestinal stromal tumors are usually submucosal or intramural, lack tactile-like corpuscles, and they express CD117 and often CD34 and actin. Neurofibromas contain in contrast with SCH heterogenous cell population which includes CD34+ and/or EMA+ cells and scattered axons, in addition to S100+ Schwann cells. Tactile-like corpuscles are common in diffuse type neurofibroma of the skin and soft tissue, but this type of neurofibroma is exceedingly rare in the gastrointestinal system[14,15]. Ganglioneuroma contains in addition to Schwann cells at least a few ganglion cells which express synaptophysin and NSE[17,22]. Mucosal neuroma, a lesion associated often with multiple endocrine neoplasia, consists of hyperplastic bundles of nerve fibers, including frequent axons[1,16,22]. Immunohistochemically, it contains S100+ Schwann cells, CD34+ endoneurial cells and EMA+ perineurial cells (like cutaneous encapsulated palisaded neuroma). Mucosal schwannoma[18,19] is well circumscribed, surrounded by dense lymphoid infiltration and it express S100 protein and GFAP. Some cases contain axons and/or CD34+ cells. It lacks tactile body like corpuscles. Mucosal colonic perineurioma is S100 protein-negative and it express perineurial cell markers, such as EMA, GLUT-1 and claudin-1[20,21].

In sum, we described mucosal SCH with focal features of tactile corpuscle-like bodies. The case further supports histogenetic relationship between mucosal SCH and tactile corpuscle-like bodies. Both lesions are benign and none of the published cases was associated with inherited syndromes.

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