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# A rare case of pseudoglandular schwannoma

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**Abstract**

A pseudoglandular schwannoma is a rare benign tumor. Controversy in the literature regarding the histogenesis of pseudoglandular schwannoma exists. Histological features of pseudoglandular schwannomas are different than those found in classical schwannomas. A patient with pseudoglandular schwannomas has a good prognosis, and no cases of recurrence have been reported in the literature.

**Keywords:** Pseudoglandular schwannoma, Pseudocyst, Schwannoma

## Introduction

Pseudoglandular schwannoma is a rare type of schwannoma first described by Ferry et al. [1] in 1988. Known variants of schwannoma are cellular, plexiform, ancient, melanocytic schwannoma, and glandular schwannomas [2]. Pseudoglandular schwannoma has not yet been included in the textbooks. Very few cases of this type of schwannoma have been reported as only 10 cases in the literature can be found. Pseudoglandular schwannoma is histologically similar to typical schwannoma findings accompanied by cystic spaces of various sizes. Although histogenesis is a controversial topic in the literature, two theories have been proposed [1, 2]. The first theory suggests that cystic cavities occur due to the degenerative change of schwannoid cells, and the second suggests that Schwann cells develop via “epithelial metaplasia”. In this study, a case of pseudoglandular schwannoma with a predominant pattern is presented. Pseudoglandular schwannoma is uncommon and can serve as a diversion when attempting to distinguish it from other tumors that are included in the differential diagnosis of schwannoma.

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**Informed Consent**
The authors stated that the written consent was obtained from the patients presented with images in the study.

**Conflict of Interest**
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## Case presentation

In our case, a 49-year-old female patient from whom informed consent was obtained to publish this case report was admitted to the orthopedic clinic with the complaint of a mass in the right shoulder. On a contrast-enhanced magnetic resonance (MR) image, a well-circumscribed, 4 x 4 x 3 cm, septated, heterogeneous mass was observed. The tumor did not invade surrounding muscle tissues and was reported to be compatible with synovial sarcoma.

Part of the material was excised for pathologic examination. The material consisted of small, brown tissue suggestive of the cyst wall on macroscopic examination. Microscopic examination under low magnification revealed tumoral tissue with an encapsulated appearance and small cystic cavities(Figure 1). When the cystic cavities were viewed under higher magnification, it was observed that 2–3 rows of epithelial-looking cells lined the lumen. These cells had large cytoplasmic areas and formed eosinophilic zones on the apical sides (Figure 2). In the lumen, eosinophilic secretions with bleeding, foamy, and hemosiderin-laden macrophages were observed in the cyst wall. It was observed that around the cysts, spindle-shaped, narrow eosinophilic cytoplasmic cells formed bundles. Peripheral nerve bundles and muscular tissues were observed within the soft tissue around the tumor. It was noticed that the peripheral nerve bundle was continuous with the tumor capsule.

Figure 1: A,B,C: Pseudoglandular areas at low magnifications. D: Foamy macrophages and hemosiderin-laden macrophages on the cyst wall (Hematoxylin and eosin [H&E] staining x40)

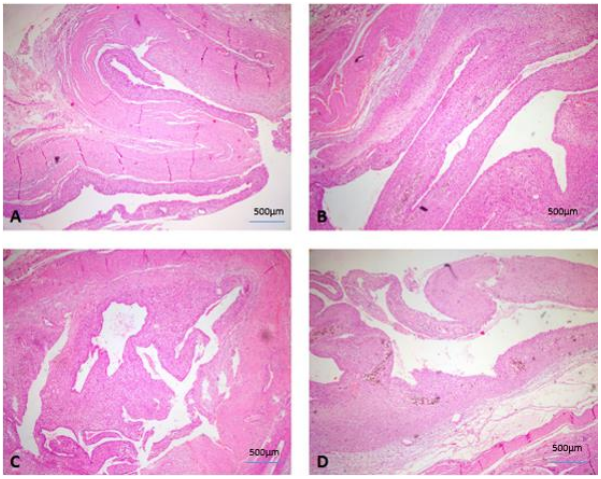
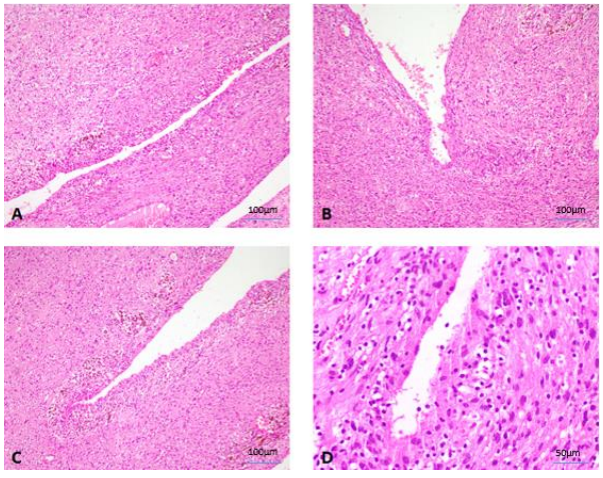
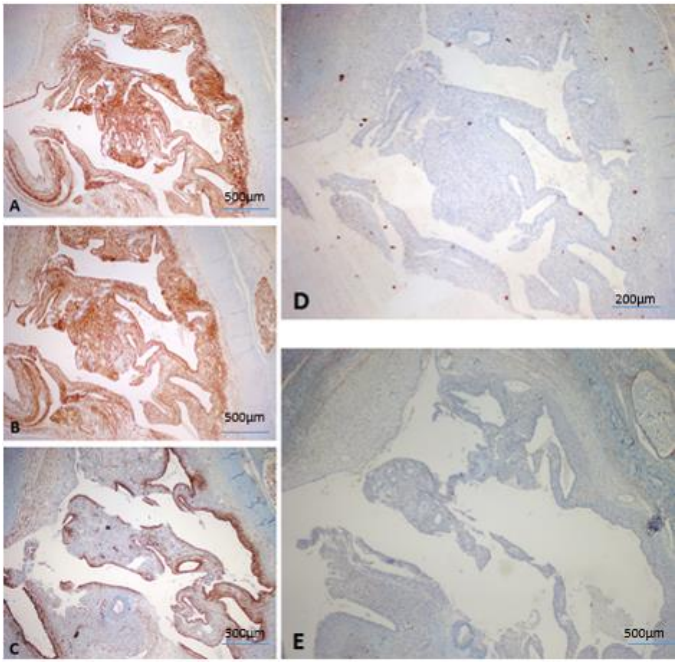



Figure 2: A, B, C, D: 2–3 rows of epithelial-looking cells lined the lumen, these cells had large cytoplasm and formed an eosinophilic zone on the apical side. (A,B,C: H&E x200 D: H&E x400)




In the immunohistochemical study, the pseudostratified epithelial cells exhibited diffuse and strong positivity for cytoplasmic S100, WT1, and CD56. Negative staining with pan-cytokeratin, TLE-1, and epithelial membrane antigen (EMA) was observed in both spindle areas and cystic cavities (Figure 3). The Ki67 proliferation index was found to be 3% in spindle cell areas of the tumor.

Figure 3: A: Strong and cytoplasmic staining with CD56 (x40) B: Strong and cytoplasmic staining with S100 (x40) C: Strong and cytoplasmic staining with WT1 (x40) D: Negativity staining with pan-cytokeratin (x100) E: Negativity staining with EMA (x40)




With the present findings, our case was diagnosed as a pseudoglandular schwannoma. After the diagnosis was made, total excision was of the mass was performed. No additional treatment was required. Complications and recurrence did not develop in the patient for whom regular follow-up was recommended.

## Discussion

A pseudoglandular schwannoma was first described in a mass with spinal canal localization by Ferry et al. [1] in 1988. In another article, a series of three cases with retrobulbar, submandibular, and shoulder locations was presented [2]. Two of these cases involved female patients, and one involved a male. Ages ranged between 24 and 37. A similar case with one shoulder location was found in the literature. Our case involved a middle-aged female patient and describes the second case of pseudoglandular schwannoma was located in the shoulder. In a retrospective study by Robinson et al. [3] in 2005, 202 schwannoma cases were examined, and 16 (7.9%) were found to contain pseudoglandular structures. While the frequency of pseudoglandular structures varies between cases, only one cystic cavity was observed in one case, and more than 80 cystic spaces were observed in another case. In a similar study with more cases, pseudoglandular structures were seen in 61 (6.3%) of 971 schwannoma cases [4]. In this study, it was reported that an average of seven cystic cavities were observed per case, and no correlation between the tumor size and the number of cystic spaces was found.

Controversy in the literature regarding the histogenesis of pseudoglandular schwannoma exists. The theory considers the capabilities of nervous system cells to differentiate into various cells. Cells that form pseudoglandular structures undergo "epithelial metaplasia". In the case report by Ruggeri [5], the findings of focal epithelial membrane antigen (EMA) and cytokeratin positivity in pseudoglandular areas support this theory. It has also been reported that pseudoglandular schwannoma may progress to glandular schwannoma [6, 7]. The second, more common theory states that pseudoglandular areas are formed by degeneration and cystic enlargement of the Verocay bodies [1, 2]. The observation of hemosiderin-laden macrophages accompanying pseudoglandular areas in most cases may also be evidence that these areas are degenerative [4]. In the case presented by Ferry et al. [1], the cells lining these cystic cavities proved to be of schwannoid origin based on results from both ultrastructural and immunohistochemical evaluations. In our case, the fact that the pseudoglandular areas did not express pancytokeratin and the diffuse and strong reaction observed with S100 supports the second theory.

When the histopathology of pseudoglandular schwannomas is examined, it can be seen that Antoni A and B areas, which cause the biphasic appearance observed in typical schwannoma morphology, are accompanied by various enlarged cystic cavities. In most cases of pseudoglandular schwannomas encountered in literature, the cystic cavities are observed as scattered foci within the tumor and do not form a dominant pattern. In the article of Deng et al. [8] in which pseudoglandular schwannoma with skin localization is presented, it was demonstrated that the dominant pattern was pseudoglandular structures. In our case, pseudoglandular areas constituted the dominant component of the tumor and created scarring of a small tumor.

In parallel with the idea that pseudoglandular areas are of schwannoid cell origin, it has been reported that these areas exhibited a diffuse and strong positivity for S100. In our case, a strong and diffuse reaction was obtained with CD56 and WT1 in addition to S100. It is known that schwannomas are frequently stained with CD56 [9, 10]. CD56 staining of a pseudoglandular schwannoma has been reported in one case in the literature [11]. However, to our knowledge, cytoplasmic staining with WT1 in pseudoglandular areas has not been previously reported. If supportive findings are obtained in future studies, WT1 may be a helpful marker in the diagnosis of pseudoglandular schwannomas.

In our case, a neurofibroma was excluded after observing strong and diffuse staining with S100. Diffuse staining with S100 is not expected in neurofibromas because neurofibromas contain a mixed cell population [11]. Another difference is that neurofibromas are infiltrative, whereas schwannomas are usually encapsulated. A malignant peripheral nerve sheath tumor was excluded based on the absence of pleomorphism and mitosis in the case and diffuse S100 staining. Contrary to what is expected in schwannomas, our case had dense cystic spaces that caused a heterogeneous appearance radiologically. In the adult patient, the diagnosis of the mass located in the vicinity of the large joint was synovial sarcoma. Synovial sarcoma was excluded because we observed TLE-1, pan-cytokeratin, and EMA negativity.

Patients with pseudoglandular schwannomas have a good prognosis, and no cases of recurrence have been reported in the literature. Removing schwannomas can cause some sensational deficit; however, this deficit is temporary [12].

**Conclusion**

A pseudoglandular schwannoma is a rare entity with controversial histogenesis. According to the common view in the literature, pseudoglandular areas are formed as a result of degenerative changes in schwannomas. The diagnostic criteria of this entity, which has not yet been found in textbooks, are not clear, and it may be possible to establish a diagnostic criterion by determining a cut-off value based on the number of pseudoglandular structures. Our case draws attention to schwannom variants and their histopathological features. The presentation of such cases can be useful for awareness of this rare structure, and pseudoglandular schwannoma cases can be considered as a different entity.

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