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Schwannoma of the tongue in a child

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INTRODUCTION

A schwannoma or neurilemmoma is a benign tumour originating from Schwann cells of the nerve sheath (*Enoz et al.*, 2006). It is a slow growing, usually solitary and encapsulated tumour (*Enoz et al.*, 2006; *Karaca et al.*, 2010). Extracranially, about 25% to 40% of all schwannomas are seen in the soft tissues of the head and neck region (*Vafiadis et al.*, 2005; *Harada et al.*, 2001; *Ma et al.*, 2010). They often originate from the VIIIth cranial nerve (*Vafiadis et al.*, 2005; *Dreher et al.*, 1997). Intraoral schwannomas are rare and account for 1% of schwannomas of the head and neck region (*Enoz et al.*, 2006; *Mevio et al.*, 2002; *Lahoz Zamarro* and *Galve Royo*, 1990). A Schwannomas may occur at any age but are more common between the 4^{th} and 6^{th} decade of life (*Baranovic et al.*, 2006). Tumours in the oral cavity are often seen in the 2nd and 3rd decade of life, but are very rare in those aged 10 years or younger (*Chiapasco et al.*, 1993; *Nakasato et al.*, 2005; *Kumar* and *Rajan*, 2004). The etiology is still unknown. The disease is generally asymptomatic and symptoms depend on size and location of the tumor (*Dreher et al.*, 1997). Recurrence is rare after complete surgical resection (*Mevio et al.*, 2002). We report a case of schwannoma in a child, involving the lateral border of the tongue. After complete intraoral surgical excision the patient has not shown recurrence during 5 years of follow-up.

CASE REPORT

We present a rare case of a 10 year-old-boy with a schwannoma on the left lateral border of the tongue. Initially he had no symptoms. His mother noticed a swelling on the left lateral border of the tongue one month before. Later, the swelling was associated with pain and tenderness while swallowing. He was examined by a paediatrician and otorhinolaryngologist in a General Hospital where the fine needle aspiration cytology (FNAC) of the lesion was performed, which was initially diagnosed as a pleomorphic adenoma. He was then referred for further treatment.

Intraoral examination revealed a well-demarcated, palpable, elastic submucosal node, 15 mm in diameter (Fig. 1). Examination of the rest of the oral cavity revealed no other lesions. There was no lymph node enlargement in the neck. The patient's medical history was unremarkable. Results of routine laboratory tests were within normal limits. We repeated the FNAC and it was inconclusive: the smears were hypocellular with few naked nuclei and scanty myxoid stroma.

Additionally, MR of the tongue and oral cavity was performed and revealed a submucosal tumour measuring 1.8 x 1 x 1.3 cm in the middle third on the left side of the tongue, which on axial T2 and coronal STIR sequences showed high signal intensity and post-contrast showed relatively good contrast uptake (Fig. 2). The tumour was well demarcated to surrounding structures of the tongue. Other structures of the tongue and oral cavity were without pathomorphological signs. There was no lymph node enlargement in the neck.

Considering equivocal FNAC finding, we decided on complete resection of this tumour. The lesion was completely excised by intraoral approach and surgical defect was closed directly (Fig. 3 and 4).

Histopathological examination of the surgical specimen revealed the typical morphological pattern of a schwannoma, composed of well differentiated Schwann cells with evident nuclear palisading throughout the tumour mass, predominantly the so-called Antoni A pattern, forming nuclear palisades, named Verocay bodies. Immunohistochemistry showed clear positive staining for S-100 protein, proving the tumour's origin (Fig. 5).

The postoperative course was uneventful. The patient has been followed up for 5 years and there has been no evidence of recurrence (Fig. 6). Mobility of the tongue is good and speech is normal.

DISCUSSION

Schwannomas, in the English literature most often called neurilemmomas, are well known in ear-nose-and-throat-medicine as benign tumours of the nerve-sheath, often in the form of acoustic nerve neuromas (*Vafiadis et al.*, 2005; *Dreher et al.*, 1997). They are seldom found in other peripheral nerves of the head (*Budde et al.*, 2001). Intraoral schwannomas are rare and account for 1% of lesions of the head and neck region (*Karaca et al.*, 2010). Among intraoral lesions, however, the tongue is a more common site, followed by, in decreasing order of frequency, cheek mucosa, the floor of the mouth, palate, lips and gingiva (*Enoz et al.*, 2006; *Lahoz Zamarro* and *Galve Royo*, 1990; *Pereira et al.*, 2008). Some authors stress that the dorsum of the tongue is the most common site (*Nakasato et al.*, 2005). We believe that the frequency of intraoral schwannomas is actually less than 1%, because this 1% includes tumours arising from the base of the tongue, which should be counted among the tumours of the oropharynx instead of the mouth. In some studies it has been noted that schwannomas may occur at any age but are more common between the 4th and 6th decades of life with a 1.6:1 female predilection, while other authors claim that the tumours have no gender preference and that the oral cavity is involved in the 2nd and 3rd decades of life (*Karaca et al.*, 2010; *Baranovic et al.*, 2006; *Chiapasco et al.*, 1993; *Kumar* and *Rajan*, 2004; *Hsu et al.*, 2006). There are only two patients of similar age in literature as the one in this report. *Pereira et al.* (2008) present a 12-year-old boy with one year follow up and *Enoz et al.* (2006) present a lingual schwannoma in a 7-year-old boy. To our best knowledge, the present case is one of two youngest patients with an intraoral schwannoma in the literature.

The major clinical sign of lingual schwannoma is a painless mass (*Hsu et al.*, 2006). The clinical differential diagnosis of lingual schwannoma includes malignant lesions such as sarcomas and benign lesions such as granular cell tumours, salivary gland tumours, leiomyomas, rhabdomyomas, lymphangiomas, haemangiomas, (epi)dermoid cysts, lipomas, inflammatory lesions and ectopic lingual thyroid (*Enoz et al.*, 2006; *Karaca et al.*, 2010; *Baranovic et al.*, 2006). *Hwang et al.* (2005) stress that a small and slow-growing mass in the tongue with a history of tongue bite is first suggestive of neurilemmoma, as well as neurofibroma, lingual cyst, and minor salivary gland tumour. Although the diagnosis of schwannoma is usually made post-operatively by histological identification, modern imaging techniques can provide useful indications. Imaging features, including CT and MRI, are relatively non-specific, but they can help in the differential diagnosis on the basis of shape and location. Magnetic resonance imaging is superior to other imaging modalities. On MRI a schwannoma is smooth and well-demarcated. This tumour is isointense compared to muscle on T1-weighted images and homogeneously hyperintense on T2-weighted images (*Flickinger et al.*, 1989). The preoperative FNAC from the swelling of the tongue could be diagnostic or inconclusive especially if repeated twice or more, because a lot of blood would contaminate the smears (*Spandow et al.*, 1999). There is a possibility to misdiagnose neurilemmoma with pleomorphic adenoma or a malignant tumour like leiomyosarcoma (*Handa et al.*, 2009; *Patnayak et al.*, 2007).

The treatment is exclusively surgical and consists of complete surgical excision (*Mevio et al.*, 2002). There are no reports of malignant transformation of schwannomas (*Mevio et al.*, 2002). Peripheral nerve sheath tumours rarely occur in the oral cavity (neurofibroma,

schwannoma, palisaded encapsulated neuroma) and the definitive diagnosis requires histological examination (*Hsu et al.*, 2006; *Go*, 2002).

Gallesio and *Berrone* (1992) indicate that after the final histological diagnosis of schwannoma, the patient needs a thorough general examination to check for the presence of other characteristic signs of von Recklinghausen's syndrome, which has a probability of malignant transformation ranging between 5 and 16%. In our opinion, this procedure is not necessary because schwannomas are part of neurofibromatosis type II (*Von Deimling* and *Perry*, 2007; *Stemmer-Rachamimov et al.*, 2007). In contrast to multiple neurofibromas (von Recklinghausen's disease), schwannomas almost never undergo malignant transformation (*Karaca et al.*, 2010; *Dreher et al.*, 1997; *Baranovic et al.*, 2006; *Güneri et al.*, 2006).

A point of note, in many papers authors have been citing that schwannomas are benign tumours originating from Schwann cells of the nerve sheath and have been identified by Virchow in 1908 (*Enoz et al.*, 2006; *Mosharrafa et al.*, 1997; *Wada et al.*, 2001). However, there is no reference to the original publication. Rudolf Ludwig Karl Virchow was born 13 October 1821 in Schivelbein, Pomerania. He was a German doctor, anthropologist, public health activist, pathologist, prehistorian, biologist and politician. He is referred as the "Father of Pathology," and founded the field of Social Medicine. He died on 5th of September, 1902 in Berlin (http://en.wikipedia.org/wiki/Rudolf_Virchow). One must ask himself, whether it is possible that Virchow identified Schwann cells in 1908, having in mind the fact that he died six years earlier? Nerve sheath tumours were first described in 1910 by Verocay (*Wippold II et al.*, 2006; *Verocay*, 1910).

CONCLUSION

The schwannoma of the tongue is extremely rare, especially in children, and, there are very few similar case reports in the present literature. Hereby we would like to add one new case of a schwannoma of the tongue as an example of a lesion which is often not taken into account during clinical practice, or even considered as a possible diagnosis.

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FIGURE LEGENDS

FIGURE (1) The patient at initial examination showing a well-demarcated, submucosal swelling on the left lateral border of the tongue, 15 mm in diameter.

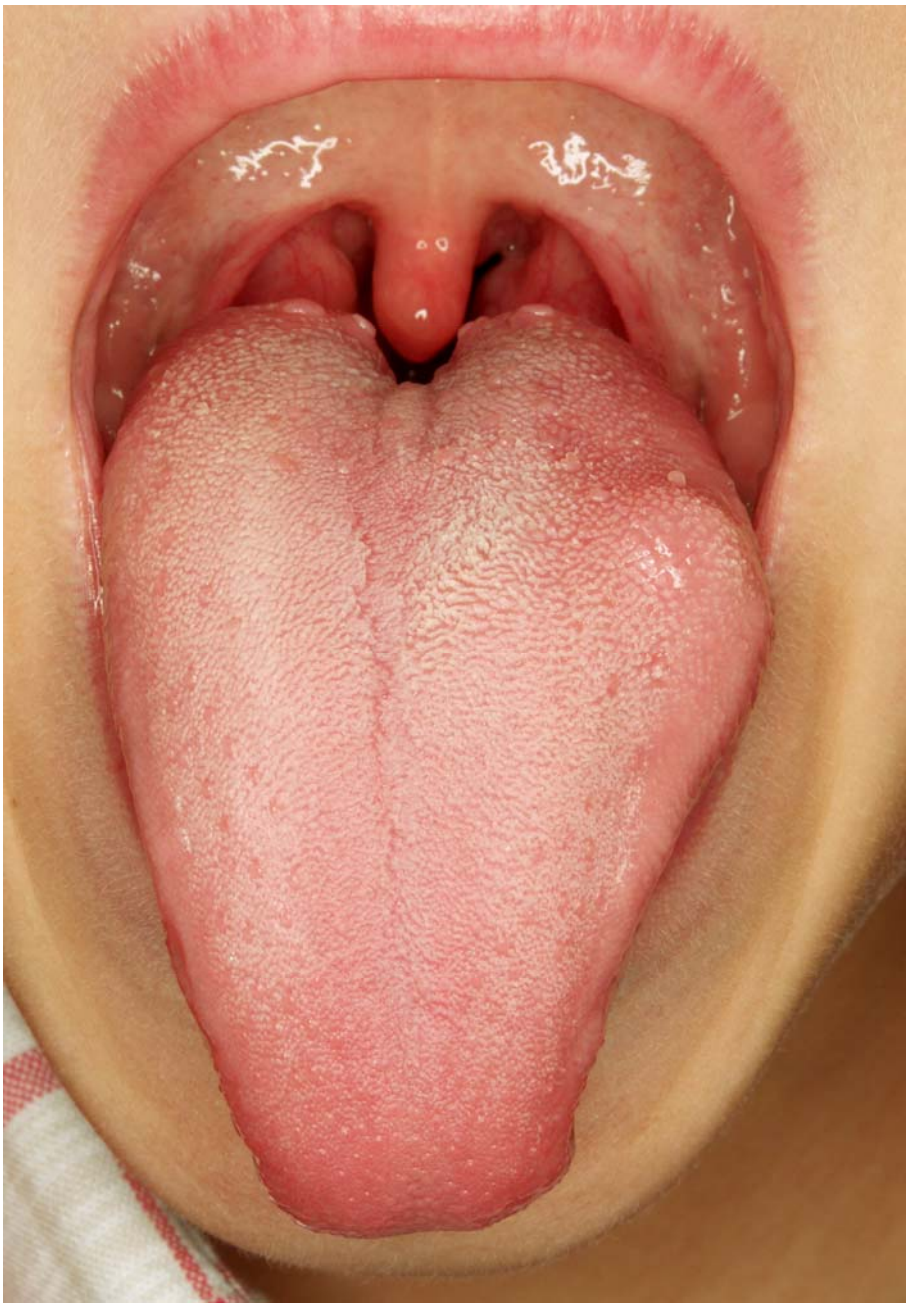

FIGURE (2) MRI coronal image (with contrast) showing a well demarcated mass, occupying the left lateral border of mobile part of the tongue.

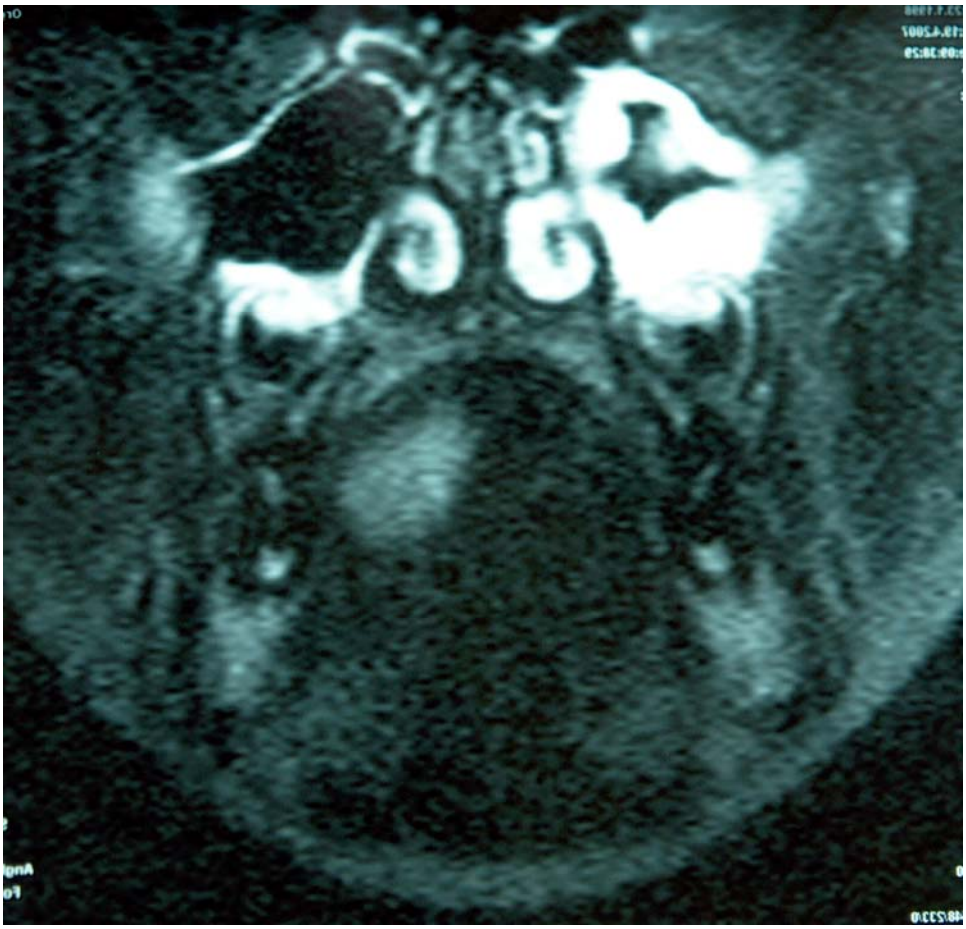

FIGURE (3) Intraoperative view after tumour excision.

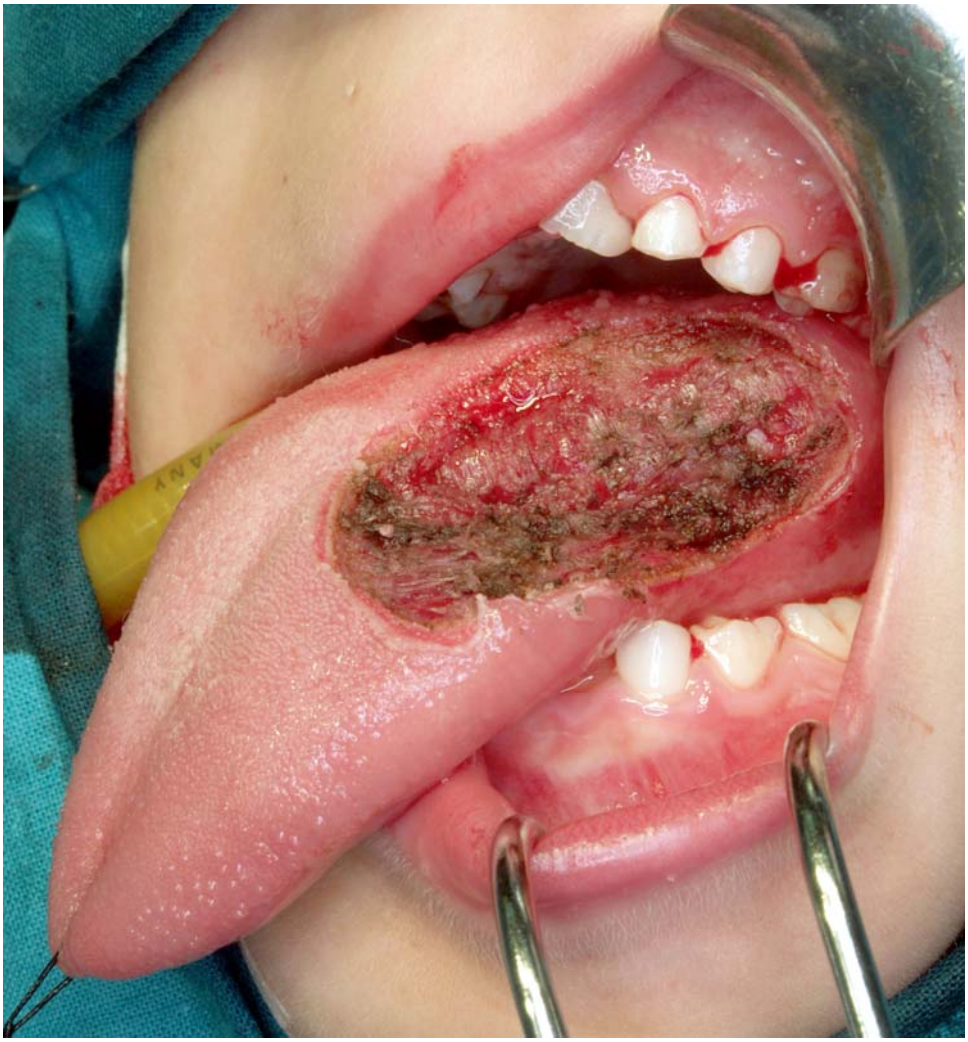

FIGURE (4) Surgical specimen including the tumour within the tongue.

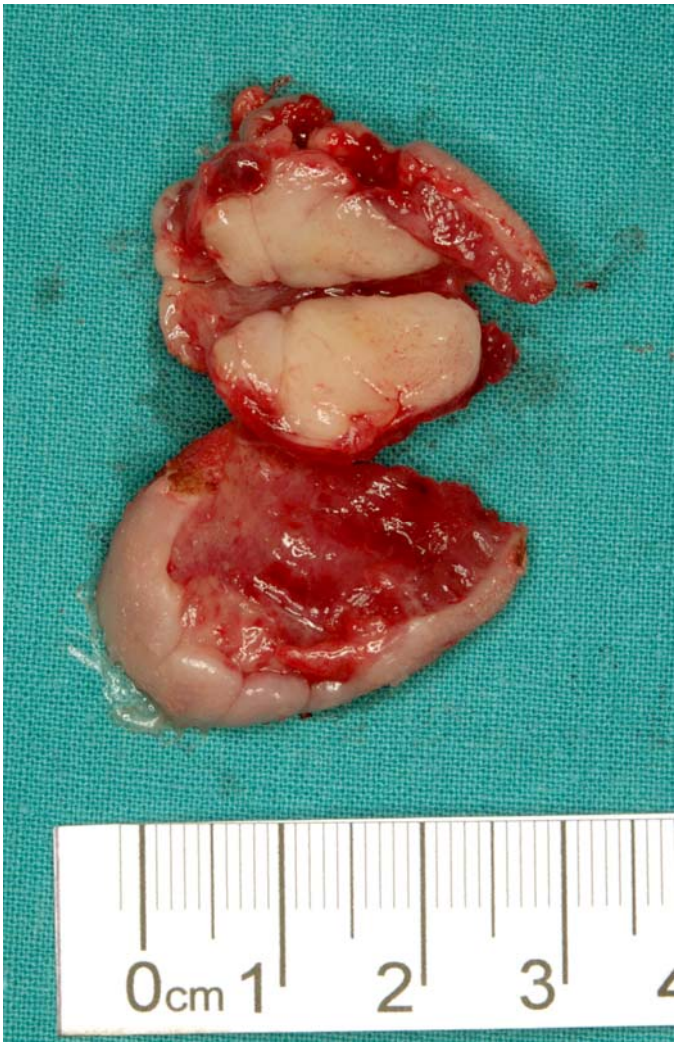

FIGURE (5) Areas of well differentiated Schwann cells with nuclear palisading throughout the tumour mass, predominantly the Antoni A pattern, forming nuclear palisades (Verocay bodies) characterized by linear arrangements of elongated tumour nuclei (haematoxylin-eosin stain, original magnification x 100).

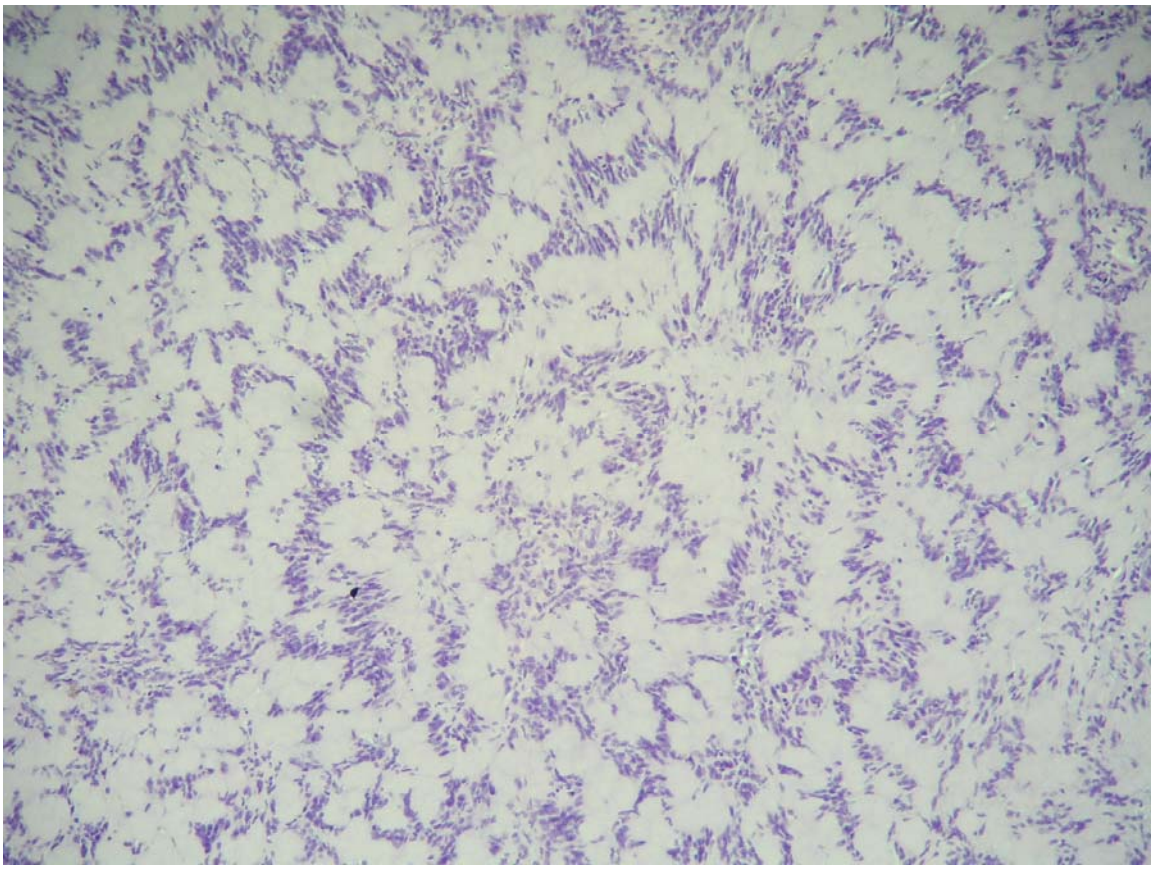

FIGURE (6) Postoperative view.