CASE REPORT

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Intraparotideal Facial Nerve Schwannoma

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ABSTRACT Schwannomas are rare, benign, slow-growing tumors of schwann cells of peripheral and cranial myelinated nerves. Facial nerve schwannomas are mostly found in intratemporal region however intraparotideal involvement of tumor is extremely rare and they are mostly diagnosed intraoperatively. In this study, 50 year-old woman suffering from a painless swelling and numbness in the right parotideal region that presented over 10 years was admitted to our clinic. After preoperative diagnostics are performed, with initial diagnosis of malignant parotideal tumor considering the size and facial nerve involvement, total parotidectomy is planned. During the operation after frozen section, it is realized that a benign tumor was originated from facial nerve and postoperatively histopathological result was schwannoma. As a result, it is important to think facial nerve schwannoma which is hard to preoperatively diagnose as a differential diagnosis of parotideal masses.

Keywords: Parotid diseases; neurilemmoma; facial nerve

Schwannomas are rarely seen, benign, well-capsulated tumors which are originated from schwann cells of myelinated peripheral nerves.[1] At craniocephalic area twenty five percent of all schwannomas are found.[2] Although vagus nerve is the most common affected cranial nerve from schwannoma, facial nerve schwannoma (FNS) can be also rarely detected. It mostly arises at the intratemporal part of the nerve. Incidence of intraparotid FNS is just 10% of all facial nerve schwannomas.[3] The most common complaint is chronic asymptomatic single-sided parotid mass. Even though the tumor is related to facial nerve; its dysfunction is seen in only 20% of all patients.[4,5] Pleomorfic adenoma, which is the most common benign tumor in parotid gland, has similar clinical features and can cause late diagnose of intraparotid FNS preoperatively. For diagnosis ultrasound, fine needle aspiration biopsy (FNAC), computerized tomography (CT) and magnetic resonance imaging (MRI) can be helpful however intraparotid FNS are usually recognized during surgery. In our study, we present our clinical approach related to the diagnosis and management of a case with intraparotid FNS.

CASE REPORT

A 50 year-old woman suffering from a painless swelling and numbness in right parotideal region that presented over 10 years was admitted to our clinic. On clinical examination, the swelling was 2×2 cm in size, painless, semi-mobile. The patient had no history of facial palsy and her facial functions were intact. She had no systemic disorders. Parotid gland ultrasound reported lobulated multicystic mass approximately 24x22 mm in size, located in deep lobe of parotid gland. Additionally blood tests were normal and FNAC result was not diagnostic. On MRI, a heterogeneously-contrasted multicytic 28×52x32 mm lesion was detected in deep lobes of right parotid gland spreading from parapharyngeal space to foramen ovale which is hypointense in T1 sections and hyperintense in T2 sections. MRI images are shown in Figure 1. Perineural inva-

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sion of the primary parotideal mass was also mentioned in the report. With pre-diagnosis of malignant parotideal tumor surgical intervention was decided. During the surgery as shown in Figure 2 after incisions facial nerve truncus and its branches were detected and preserved. In parapharyingeal region reddish bright well-encapsulated, multilobular, surrounding facial nerve tumor was dissected. For its unusual localization and presence, frozen section was performed and reported benign. We dissected the tumor from the facial nerve using microscope magnification to preserve as many fibers as possible. Afterwards we performed surgical excision as a neural unsheathing and the mass was excised totally with superficial and deep lobe of parotid gland. After the operation, we verified neural functions of frontal, zygomatic, buccal and marginal branches of facial nerve by a needle neurostimulator as intact and unharmed. Although postoperatively first days, patient had grade 4-5 facial palsy (House-Brackmann classification), after 3-month follow-up it was degraded to grade 3.

Histopathologically, tumor stained routinely hematoxylin and eosin (H&E) and spindle-type cells were detected. Both Antoni A and B areas were found as shown in Figure 3. Immunohistochemical tests confirmed that neoplastic cells were EMA negative and S-100 positive. Ki-67 proliferation index was 1-2%. Regarding to these findings, the histopathological result was reported as schwannoma. For publication we received written informed consent from the patient.

DISCUSSION

Ibarz first discovered the pathologic findings of IFNS in 1927. Afterwards in 1949, O'Keefe published the first complete case report on the diagnosis and management of IFNS.[4] Since it is a rare condition, it is really challenging to distinguish preoperatively and treatment management is debatable. Most of FNS are found in intratemporal region only 9% of cases show intraparotideal involvement.[2] Caughey et al. studied 3,722 schwannoma diagnosed patients retrospectively and intraparotideal FNS are found in only 8 cases.[6]

Macroscopically schwannomas are encapsulated, soft, yellow-white colored tumors. As presented in our case, they can be also cystic and multilobulated. Histopathologically two tissue types are defined to characterize schwannoma, which are Antoni A and B areas.

Schwann cells founded in Antoni A area is spindle-shaped and their nuclei are arranged in palisading pattern which is called as Verocay bodies. On the other hand, Antoni B area has more cellular pleomorphism and no obvious palisading nuclei.[2] S-100 staining is used to prove neural structures of the tumor and smooth muscle actin (SMA) staining is done to exclude possible leimyoma.[2,3] For our case, the tumor was reported S-100 positive and SMA negative. No sign of necrosis, atypia or mitosis was detected in the surgical sample.

Diagnosis of intraparotideal FNS is challenging preoperatively due to its lack of incidence. Because

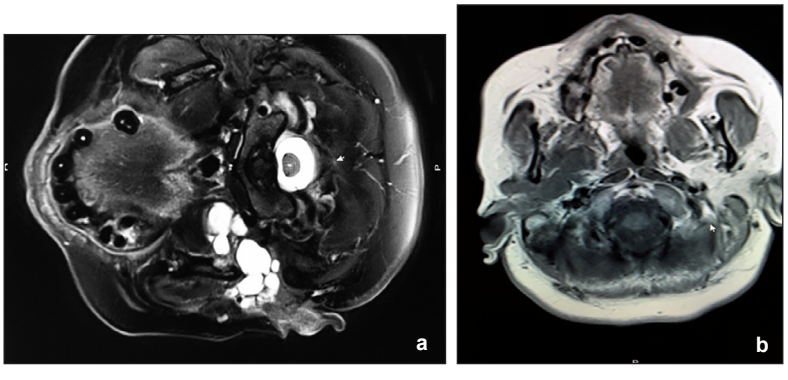

FIGURE 1: A) multicystic mass hyperintense in axial T2 sections involving deep lobe of parotis gland, **B)** multilobulated mass heterogeneously-contrasted in axial T1 section.

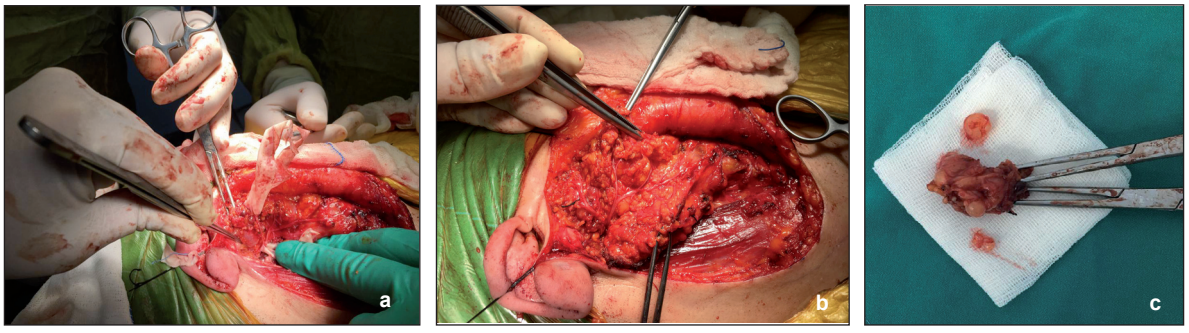

FIGURE 2: A) localization of tumor while facial nerve hanged, **B)** preserved facial nerve truncus and its branches, **C)** specimen.

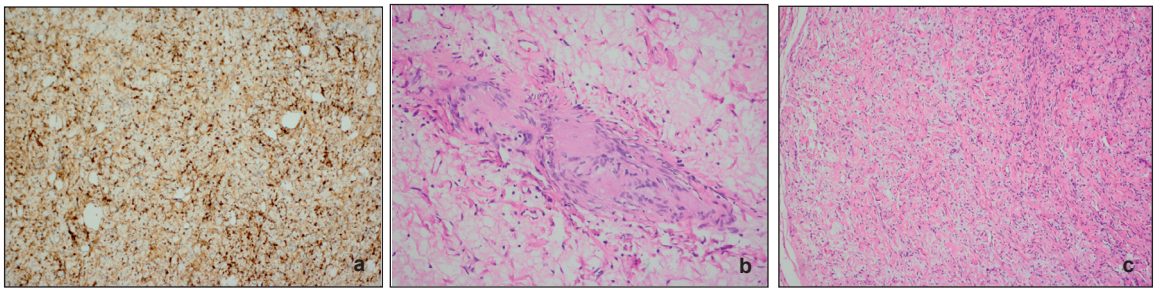

FIGURE 3: A) The lesion had positive reaction with S-100 in immunohistochemical examination X100, **B)** palisating schwann cells stained H&E X200, **C)** Antoni A and Antoni B areas H&E X100.

postoperative damage of facial nerve is quite often, it is however important. Unfortunately it is hard to distinguish radiologically from other parotideal tumors and most likely confused with pleomorfic adenoma due its resemblance. Isointens to muscle in T1 sections and well-bordered in T2 sections at MRI are general features of the tumor. They are usually heterogeneously-contrasted as a result of more cellular Antoni A areas.[3] This image on MRI is described as "target sign" according to some authors. Banks analyzed the target sign and concluded that it is inadequate to distinguish a benign from malignant one.[3,7] FNAC is a helpful tool for preoperative diagnostics and preferred due to its minimal risks and it can distinguish benign and malign masses. However its accuracy is reported 22-33% in literature.[4] In our case, cytology report resulted nondiagnostic.

Therapy of choice is still surgical excision but there are also other modalities defined in literature such as observation, partial resection or resection of tumor with sacrificing facial nerve additionally cable grafting.[4,8,9] Herein preoperative facial function, tumor localization and size must be taken into consideration. Especially during surgery, if decided, frozen section study is strongly recommended. It can differentiate benign from malign lesions and even indicate schwannoma by reporting as mesenchymal tumor, therefore it can guide the surgeon.[2] We also ran frozen section study and resulted benign. Marchioni et al. categorized intraparotideal FNS into four types according to its relation with facial nerve. In Type A, surgery can be performed without damaging the facial nerve. Type B tumors are more attached to the nerve therefore partial sacrifice of peripheral branches of facial nerve or their distal divisions may be necessary. In type C, main trunk must be sacrificed due to tumoral involvement and type D tumors require sacrificing the trunk and its main divisions to be resected.[2,8] In our case, the tumor was assumed as type A and resection was performed like stripping surgery without damaging the nerve. Intact facial nerve function is proven by intraoperative electrophysiology.

In conclusion, the preoperative diagnosis of intraparotid FNS is challenging. If it is recognized preope-

ratively with radiodiagnostics or FNAC and nerve functions were preserved, close follow-up is also recommended after informing the patient because of slow growth of the tumor. However, in most cases these tumors are identified intraoperatively and ideal treatment is surgically total excision of the tumor without damaging facial nerve.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

Idea/Concept: *Zeynep Kaptan;* ***Design:*** *Akif Sinan Bilgen;* ***Control/Supervision:*** *Zeynep Kaptan;* ***Data Collection and/or Processing:*** *Kübra Başarır, Onur Erçelik;* ***Analysis and/or Interpretation:*** *Rahmi Kılıç;* ***Literature Review:*** *Onur Erçelik;* ***Writing the Article:*** *Zeynep Kaptan;* ***Critical Review:*** *Rahmi Kılıç;* ***References and Fundings:*** *Rahmi Kılıç;* ***Materials:*** *Akif Sinan Bilgen.*

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