A case report of the first ganglioneuroblastoma of the parotid gland of an the adult

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The ganglioneuroblastoma is a rare form of pediatric tumor, involving innerved tissues of the nervous sympathetic system, whose evolution is very hard to foresee.
A 38 year-old patient whose histologic exam unexpectedly showed ganglioneuroblastoma, underwent a parotidectomy. Three years after surgery the patient is in optimal clinical conditions, without any radiological signs or clinical relapses.

KEY WORDS: Ganglioneuroblastoma, Parotid gland, Parodidectomy

Introduction

The ganglioneuroblastoma is a rare form of pediatric tumor, involving innerved tissues of the nervous sympathetic system, whose evolution is very hard to foresee [1]. The average age of diagnosis is around 22 months with a slight prevalence for male patients (ratio 1, 2:1) compared to female patients. The most common localizations are adrenergic glands (35%), retroperitoneal paravertebral ganglia (30-35%), mediastinum posterior (20%), head and neck (1-5%) and pelvis (2-3%); less frequently the tumor develops in the thymus, lungs, kidneys, anterior mediastinum or along the cauda equine [2]. Current literature only describes one case of neuroblastoma of the parotid gland, diagnosed after a post-operative histologic exam, regarding a two-year old Japanese boy [3]. However, literature mentions less than 40 cases with the localization mainly affecting the retroperitoneal region [4]. Up until now an adult case of ganglioneuroblastoma of the parotid has never been mentioned.
Almost 1% of these tumors are capable of producing metastasis that generally spread hematogenously or through the lymphatic system even though they present unpredictable characteristics: cases of spontaneous regression of neoplastic lesion were also registered [5].
The following case report describes the case of a 38-year old affected by ganglioneuroblastoma of the left parotid gland.

Case Report

A 38-year old man in normal physical and mental conditions with a case history presenting solely positivity for HCV, raised our eyebrows for the clinic discovery of a mass in the left parotid region, also asymptomatic.
The patient carried out a sonogram as a first-level examination in a different location (Fig. 1), which showed the presence of an unrefined oval formation on the left parotid side of size 38x22mm, unevenly hypoechoic, with small spots of vascular contextual signals. Below that for-

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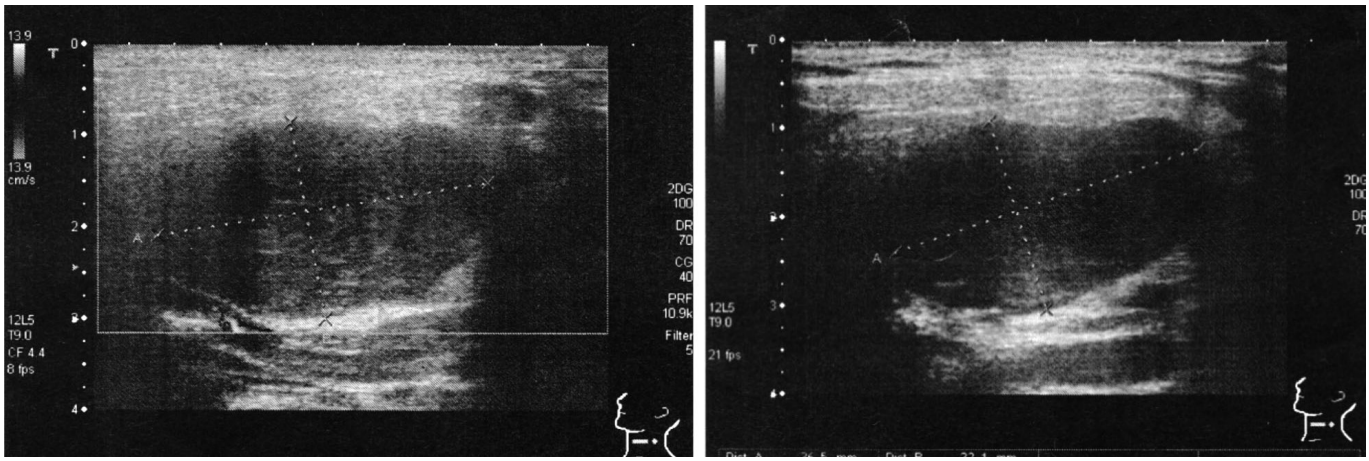


Fig. 1: Ultrasound study performed with a linear probe, noticeable is the hypoechoic formation with vascular spots.

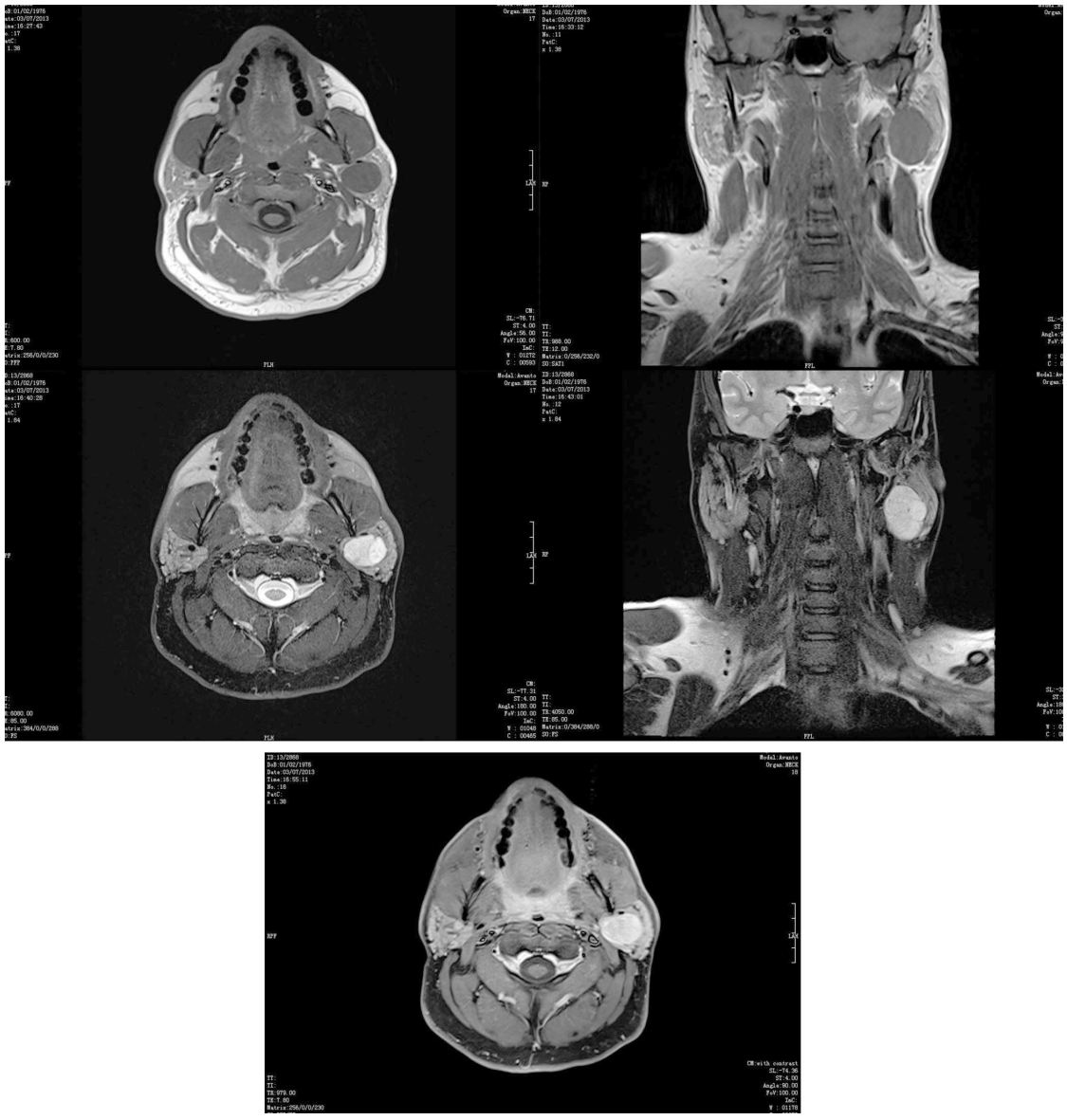
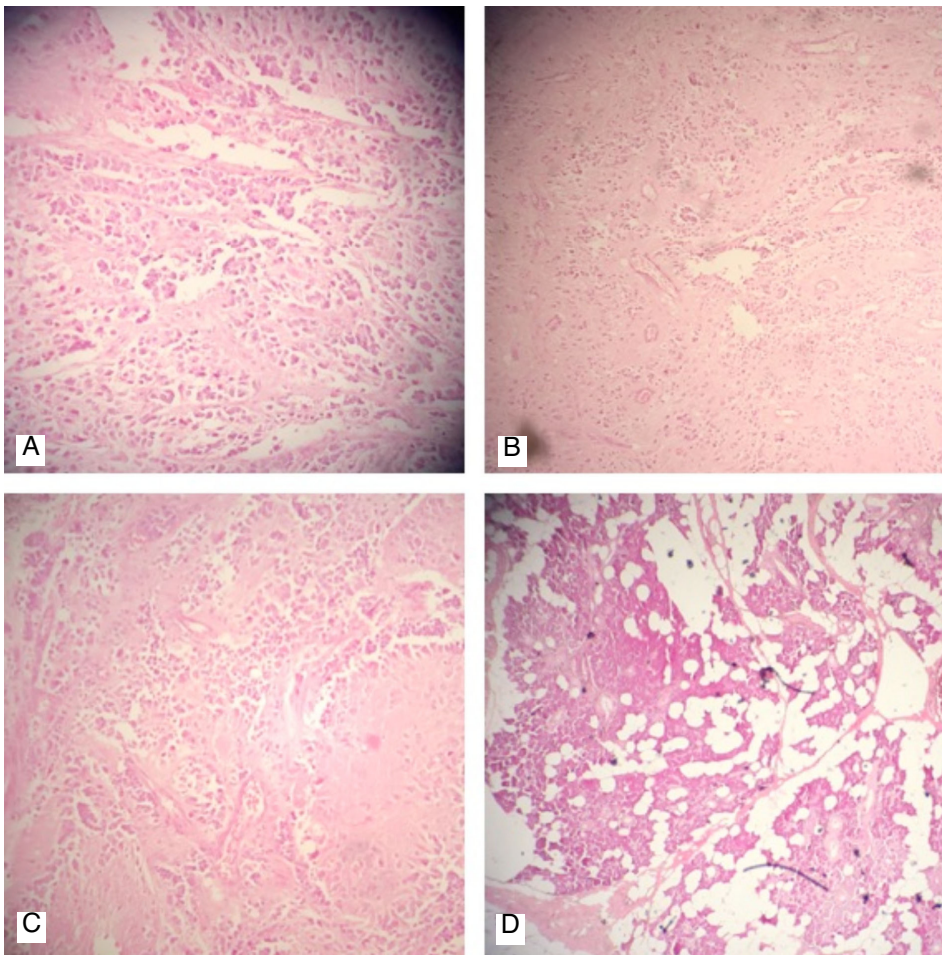


Fig. 2: RMN sequences with and without mdc, noticeable are the retromandibular vein compression and the bilateral lymphadenopathy.

Table I - *Immunohistochemical patterns*

Tissues	Immunochemistry
Healthy salivary tissue	
Healthy lymph glandular tissue	
Ganglioneuroblastoma	S-100 +
	NSE +
	Chromogranine-
	Moderate mitotic index (Ki67+)
	Non-stromal invasion

mation passed an arterial branch with bilateral lymphadenopathy, greater to the left (19 and 15 mm), but with a regular right parotid.
Given the patient's clinical conditions, he underwent a RMN with and without mdc (Fig. 2), as a second-level examination. This examination showed the presence of an expansive formation on the left parotid side with uneven hyperintensity in T2 weighted sequences and with intense impregnation of contrast effect following mdc. The 29x20x37mm lesion intersected vascular formations with high-influx of arterial pertinence originating from the external carotid (superficial temporal artery), while the retromandibular vein was not discernable because of the compressing effect carried out by the formation. Bilaterally, the lateral cervical region presented lymphonodes of maximum size of 23 mm.
The radiologist believed that such clinical conditions could be symptomatic of pleomorphic adenoma of the parotid and therefore the patient underwent surgery. After entering on the left preauricular region with a cut from the mandibular angle of the tragus and seeking the cartilaginous bridge of the digastric muscle, the facial nerve and its branches were followed through their course.
The mass occupied the retroneural portion of the gland, leading to the undergoing of the total parotidectomy. As anticipated in the preoperative imaging, the mass was delocalizing the external carotid artery and crushing at the back the posterior facial vein.
The postoperative course was not characterized by critical situation and the patient was therefore dismissed in 2° G.P.O, excluding facial nerves damages.
The histologic exam conducted on the operative piece macroscopically describes the 4x3x2 cm nodular mass with multiple necrotic-hemorrhagic areas. Furthermore it was microscopically observed the presence, in clusters separated from the well-vascularized stroma, of small, rounded cells, surrounded by pseudocapsules and with



ANATOMOPATHOLOGICAL PANELS

Figs. 3: A; B; C) Noticeable are ialinosis areas referred to tubular structures, surrounded by elements with atypical nuclei with constant nucleoli presence. Roses-like formations are also noticeable. Absence of mature Schwann cells; D) Parotid normal gland. It is possible to identify the tubular and acinosa structure of the gland.

little cytoplasm. The immunohistochemistry performed in order to better identify mass, shows the patterns illustrated in Table I.

THE PATIENT DID NOT UNDERGO ANY ADJUVANT THERAPY

All the postoperative clinical examinations, including the TC total body conducted around two months and a one year after surgery, did not show signals of recurrence of the disease.
Three years after surgery, follow-up tests show no signs of relapse of the disease

Discussion

The ganglioneuroblastoma is a tumorous disease, uncommon for children and quite rare for adults. In 2003 Koike et al. counted 37 cases [6]. The survival of the stages with R1/R2-resection borders was calculated in circa 24 months but nothing has been shown in literature in relation to the survival of R0-resections [7].
Two systems have been used relatively to the classification: Shimada System, where the prognosis is calculated (Tables II, III) and International Neuroblastoma Staging System [8] (Table IV).
This case study is the only adult case of ganglioneuroblastoma of the parotid that has ever been mentioned in current literature.
The peculiarity of the formation, subject of this study, lies in the histologic benchmarks, which turned out to be the same related to the two-year old Japanese boy affected by neuroblastoma of the parotid (positivity to vimentin, subunit alfa and beta of the S-100 protein, enolase neuro-sensitive NSE, P substance, met-enkephalin and chromogranine)
The diagnostic hypothesis over the origin of such neoplasm could be, given that the nervous and glandular tissues are both ectodermic, that in the parotid of our patient coexisted both histologies in a completely asymptomatic manner until neoplastic derivation in the nervous cellular line during adulthood. It is not possible to identify the reasons that justify these cells to cause neoplasm during adulthood.
Currently and according to the revision of the international literature about the ganglioneuroblastoma it is possible to affirm that surgery results the only viable therapeutic option [9]. A R0 resection could guarantee good outcomes in the follow-up tests of these patients.

Table II - *Shimada System*

– Stroma-poor neuroblastomas with neuroblasts that are undifferentiated, differentiating, or differentiated.
– Stroma-rich ganglioneuroblastomas with differentiated and undifferentiated neuroblasts.
– Stroma-dominant ganglioneuromas with maturing neuroblasts and mature ganglion cells.
– Composite nodular ganglioneuroblastomas with stroma-rich/stroma-dominant and stroma-poor cells and neuroblasts with various degrees of differentiation.

Table III - *Prognostic of neuroblastic tumors classified according to Shimada System*

International Neuroblastoma Classification	Patient Age (yrs)	Histology	Prognosis
Neuroblastoma (stroma-poor)	<1.5	Poorly differentiated or differentiated & low or intermediate MKI tumor	Favorable
	1.5-5	Differentiated & low-MKI tumor	Favorable
	<1.5	Undifferentiated low-MKI tumor	Unfavorable
	1.5-5	Undifferentiated or poorly differentiated tumor (regardless of MKI)	Unfavorable
	>5	All Tumors	Unfavorable
Intermixed ganglioneuroblastoma (stroma-rich)	All Ages	All Tumors	Favorable
Ganglioneuroma (maturing or mature; stroma-dominant)	All Ages	All Tumors	Favorable
Ganglioneuroblastoma, nodular (stroma-rich, stroma-poor, and stroma-dominant)	All Ages	All Tumors	Unfavorable

Table IV - *International neuroblastoma staging system*

Stage 1:
Localized tumor with complete gross excision, no gross or microscopic residual disease, and negative representative lymph nodes

Stage 2:
2A. Tumor with incomplete gross excision and negative representative ipsilateral lymph nodes.
2B. Tumor with incomplete gross excision, positive ipsilateral lymph nodes, and negative controlateral lymph nodes.
(Stage 1 + Stage 2A + Stage 2B = 25%)

Stage 3:
Unresectable tumor infiltrating across the midline, with or without regional lymph node involvement; or midline tumor with bilateral extension consisting of infiltration or lymph node involvement.

Stage 4:
Any primary tumor with dissemination to distant lymph nodes, bone, bone marrow, liver, or skin.
(STAGE 3 + **Stage 4 = 65%)**

Stage 4S
("**special" - 10%):** Localized primary tumor (as in stage 1, 2A, or 2B) with dissemination limited to skin, liver, and/or bone marrow (no bone involvement) in infants younger than 1 year. These infants have good adrenal function and more favorable prognoses.

Riassunto

Il ganglioneuroblastoma è un raro tumore che colpisce prevalentemente i giovani e raramente gli adulti.
La sua localizzazzione più frequente è a livello surrenalico e paravertebrale.
Viene descritto il primo caso di localizzazione parotidea di un ganglioneuroblastoma in un adulto sottoposto a intervento di parotidectomia totale per una neoformazione in rapida crescita.
Il paziente , a 3 anni dall'intervento è in buona salute e libero dalla malattia

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