ISSN: 2977-0033

Case Report

Journal of Clinical Research and Case Studies

Extragonadal Cervico-Parotid Yolk Sac Tumor with a Sarcoma-like Presentation in a Child: A Case Report with Fulminant Course and Literature Review

Dr Zineb Berdi*, Dr Zakaria EL Hafi, Pr Razika Bencheikh, Pr Mohamed Anass Benbouzid and Pr Leila Essakalli

Department of ENT and Head and Neck Surgery – Specialty Hospital / Ibn Sina University Hospital, Rabat, Morocco; Faculty of Medicine and Pharmacy / Mohammed V University, Rabat, Morocco

***Corresponding author**

Dr Zineb Berdi, Department of ENT and Head and Neck Surgery – Specialty Hospital / Ibn Sina University Hospital, Rabat, Morocco; Faculty of Medicine and Pharmacy / Mohammed V University, Rabat, Morocco

Received: July 24, 2026; **Accepted:** August 03, 2026; **Published:** August 10, 2026

ABSTRACT

Background: Extragonadal malignant germ cell tumors of the head and neck are exceptional in children. Among them, pure yolk sac tumor arising in the parotid region and deep neck spaces is extremely rare, and its clinical and radiological presentation may mimic a soft-tissue sarcoma, exposing the patient to diagnostic delay.

Case Presentation: We report the case of a previously healthy 5-year-old girl admitted for a rapidly enlarging right lateral cervical swelling evolving over two months. Examination found a firm, polylobulated mass with intraoral extension narrowing the oropharyngeal lumen. Contrast-enhanced CT and MRI showed a large heterogeneous mass centered on the right parotid region, extensively infiltrating the nasopharynx, tongue base, floor of mouth, parapharyngeal, masticator, submandibular and visceral neck spaces, eroding the thyroid cartilage and mandibular ramus, and associated with ipsilateral cervical lymphadenopathy; imaging findings were first suggestive of a sarcomatous tumor. Laboratory work-up showed a markedly elevated alpha-fetoprotein (AFP) of 31,087 IU/l, LDH of 695 IU/l, and normal beta-HCG. Histopathology and immunohistochemistry (positive AFP, SALL4, and focal PLAP; negative OCT3/4 and CD30; Schiller-Duval bodies) confirmed a pure yolk sac tumor. The patient was referred to pediatric oncology but died within a few days from rapid clinical deterioration, before chemotherapy could be initiated.

Discussion: This case illustrates the diagnostic difficulty of this rare tumor, whose imaging features are non-specific and can mimic sarcoma, and underscores the central role of AFP as a diagnostic clue. It also illustrates the potentially fulminant course of locally advanced disease, requiring urgent multidisciplinary management from the time of clinical suspicion.

Conclusion: In any pediatric head and neck mass with rapid growth and an aggressive imaging pattern, systematic AFP measurement should be part of the first-line work-up alongside biopsy, in order to avoid missing an extragonadal yolk sac tumor and to shorten the time to oncologic treatment.

Keywords: Yolk Sac Tumor, Endodermal Sinus Tumor, Alpha-Fetoprotein, Extragonadal, Head and Neck, Child, Parotid Gland

Introduction

Malignant germ cell tumors account for only a small proportion of pediatric cancers, but yolk sac tumor, also known as endodermal sinus tumor, represents their most frequent malignant subtype [5]. These tumors arise from primordial germ cells and most commonly occur at gonadal sites (testis, ovary); however, nearly half of cases are extragonadal, arising preferentially along the embryonic midline pathway of germ cell migration, most commonly in the sacrococcygeal region, mediastinum, retroperitoneum, and central nervous system [5].

Head and neck involvement is exceptional and represents only a minority of extragonadal forms [1,5]. The literature reports only small case series or isolated case reports, described in the orbit, nasal cavity, thyroid gland, oral cavity, ear, upper lip, parapharyngeal space, and skull base, most often occurring in infants or young children, occasionally in the setting of Aicardi syndrome or in association with a teratomatous component [2-4,6,8,9].

Alpha-fetoprotein (AFP), secreted by tumor cells in a manner analogous to the fetal yolk sac, is the reference biological marker for diagnostic orientation and disease monitoring [10]. Given the rarity of this tumor, its imaging appearance remains non-specific

Citation: Dr Zineb Berdi, Dr Zakaria EL Hafi, Pr Razika Bencheikh, Pr Mohamed Anass Benbouzid, Pr Leila Essakalli. Extragonadal Cervico-Parotid Yolk Sac Tumor with a Sarcoma-like Presentation in a Child: A Case Report with Fulminant Course and Literature Review. J Clin Res Case Stud. 2026. 4(3): 1-5. DOI: doi.org/10.61440/JCRCS.2026.v4.100

and can be mistaken for a soft-tissue sarcoma, particularly rhabdomyosarcoma, which is by far the most common pediatric soft-tissue malignancy of the head and neck. Histological and immunohistochemical confirmation is therefore essential.

We report a pediatric case of extragonadal cervico-parotid yolk sac tumor with major locoregional extension, whose imaging appearance was pseudo-sarcomatous and whose clinical course was fulminant, in order to raise awareness among otolaryngologists and pediatricians of this rare entity and of the value of systematic AFP measurement.

Case Presentation

A previously healthy 5-year-old girl, with no relevant past medical history, presented with a twomonth history of a rapidly enlarging right lateral cervical swelling, with no reported fever or general deterioration at admission.

Physical examination revealed a firm, polylobulated right lateral cervical mass with intraoral extension narrowing the oropharyngeal lumen.

Contrast-enhanced cervico-facial computed tomography (CT) showed a large, expansile, tissuedensity right lateral cervical mass, round in shape, with bosselated and irregular contours and heterogeneously enhancing after contrast injection, measuring 65×64×69 mm. Topographically, the mass infiltrated the nasopharynx superiorly and the prevertebral spaces posteriorly; anteriorly it invaded the tongue base and floor of mouth; medially it infiltrated the parapharyngeal, masticator, submandibular, parotid, and visceral neck spaces, narrowing the oropharyngeal lumen and partially filling the laryngeal lumen. It eroded the thyroid cartilage and the right mandibular ramus. Inferiorly, it reached and, in places, infiltrated the sternocleidomastoid muscle without a preserved fat plane, and encased the carotid space, which remained patent, with extension to the hypopharynx. Multiple right lateral cervical lymph nodes were present, the largest located in the right level Ia (16 mm short axis) and level Ib (10 mm short axis) chains.

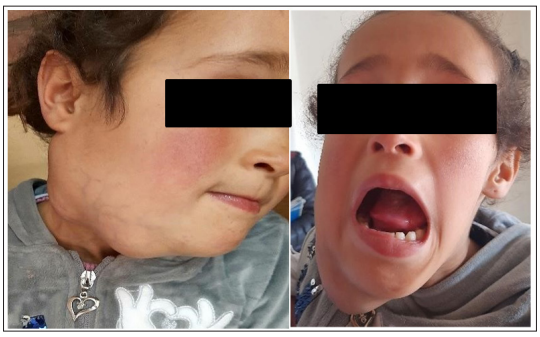

Figure 1: Image of polylobulated right lateral cervical mass with intraoral extension

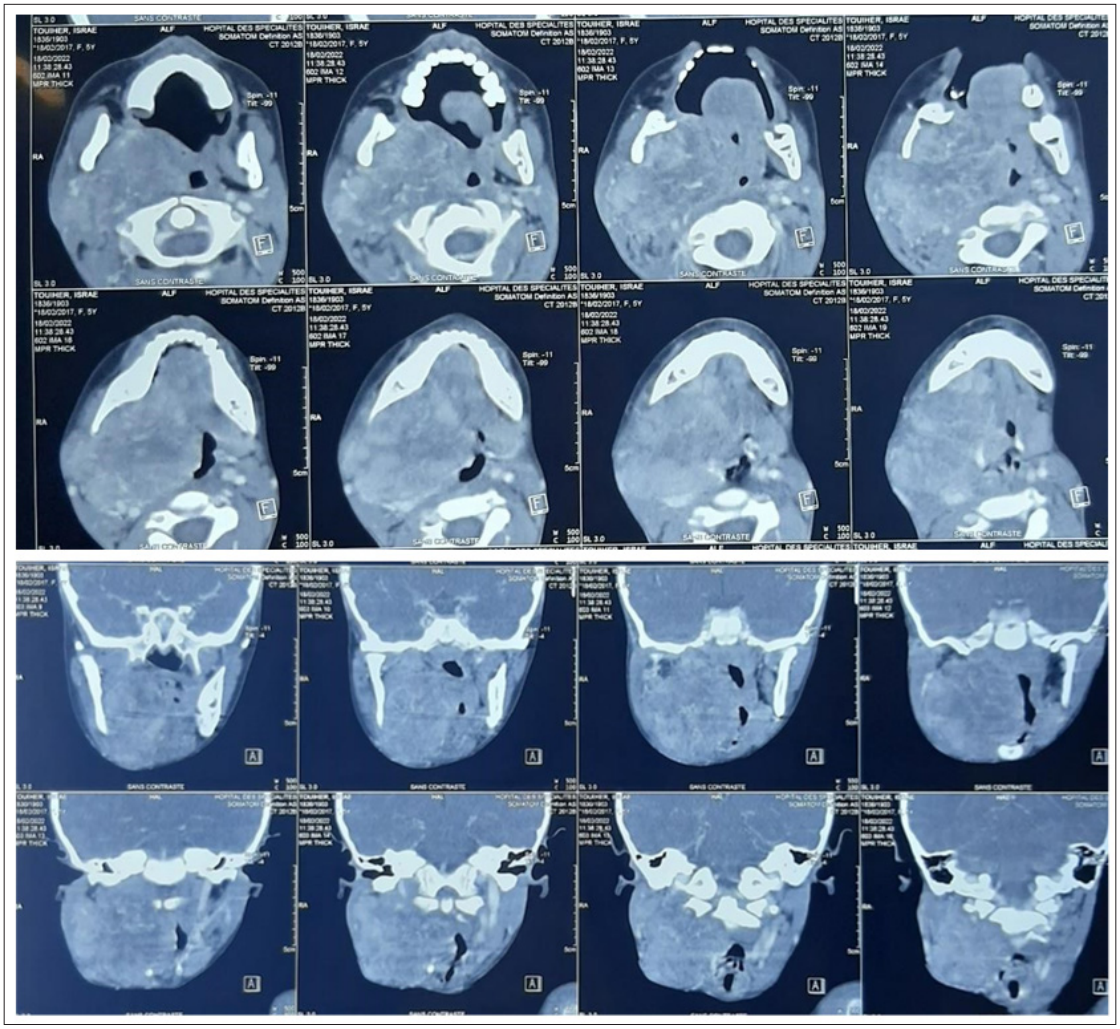

Figure 2: Cervico-facial computed tomography (CT) showed a large, expansile, tissue-density right lateral cervical mass, round in shape, with bosselated and irregular contours and heterogeneously enhancing after contrast injection

Magnetic resonance imaging (MRI), performed as a complementary study, showed a large right lateral cervical mass centered on the parotid gland, roughly ovoid, well-defined, with lobulated contours, isointense on T1-weighted images and hyperintense on T2-weighted images, containing flow-void vascular structures supplied by the superficial temporal artery, with mild diffusion restriction. After contrast administration, the mass showed progressive enhancement, initially peripheral at the arterial phase, with subsequent filling and homogenization on delayed images, measuring 63×75×76 mm. Regarding locoregional extension, MRI confirmed the CT findings, showing the same infiltration of the nasopharynx, prevertebral spaces, tongue base, floor of mouth, parapharyngeal, masticator, submandibular, parotid, and visceral neck spaces, as well as encasement of the patent carotid space and extension to the hypopharynx, better characterized owing to the superior soft-tissue contrast of MRI. The impression favored a probable right parotid mass causing oropharyngeal luminal narrowing, associated with ipsilateral lateral cervical lymphadenopathy, suggestive of a sarcomatous tumor.

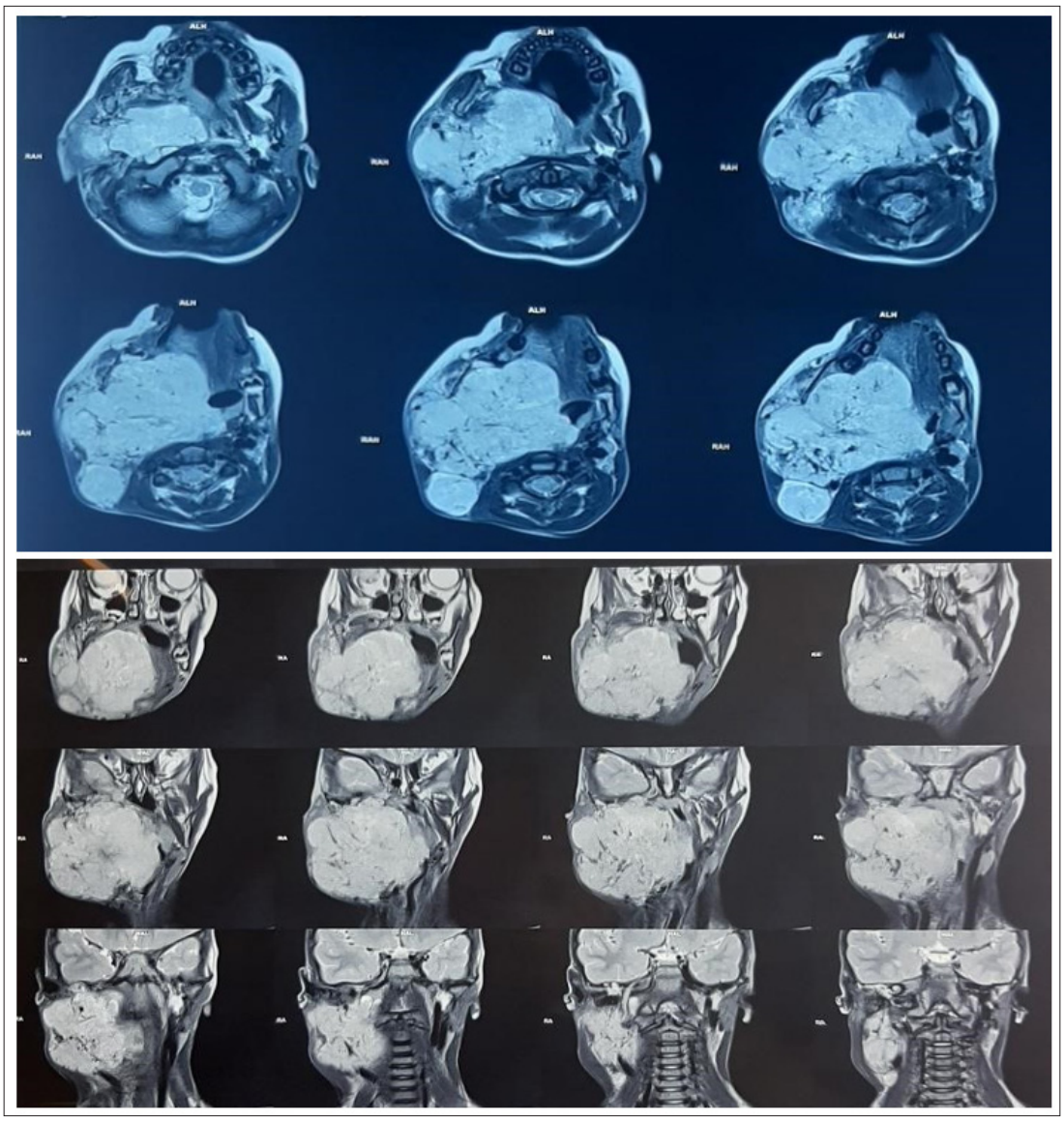

Figure 3: Magnetic resonance imaging (MRI) showing a large right lateral cervical mass centered on the parotid gland, roughly ovoid, well-defined, with lobulated contours, isointense on T1-weighted images and hyperintense on T2-weighted images

Laboratory work-up showed LDH of 695 IU/l, a markedly elevated AFP of 31,087 IU/l, and beta-HCG below 2 mIU/ml.

A biopsy of the mass was performed. Microscopic examination showed a malignant tumor proliferation composed of large cells with clear to eosinophilic cytoplasm, round nuclei with more or less irregular contours, anisokaryotic, with vesicular chromatin and a visible eosinophilic nucleolus. These cells were cohesive, arranged in a solid or reticular architecture, forming numerous SchillerDuval bodies, within a sparse, richly vascularized stroma, with marked mitotic activity. Immunohistochemistry showed strong positivity for AE1/AE3, focal membranous positivity for PLAP, negativity for CD30, negativity for OCT3/4, strong focal positivity for AFP (with weak membranous positivity elsewhere), and strong positivity for SALL4. This morphological and immunohistochemical pattern was consistent with yolk sac tumor.

The patient was referred to the pediatric hematology-oncology department. While hospitalized for planned chemotherapy initiation, she developed rapid clinical deterioration and died within a few days, before specific treatment could be started.

Discussion

Epidemiology and Anatomical Sites

Malignant germ cell tumors account for approximately 3% of pediatric cancers [5]. Yolk sac tumor is their most common malignant subtype, occurring either in pure form or as a component of a mixed germ cell tumor. Nearly half of cases are extragonadal, with preferential sites including the retroperitoneum, mediastinum, sacrococcygeal region, and central nervous system, reflecting the embryonic migratory pathway of primordial germ cells along the midline [5]. Head and neck localization remains exceptional: orbital forms number no more than about fifteen published cases, and all pediatric sinonasal and orbital locations combined amount to roughly twenty cases in the literature [1]. Isolated cases have also been reported in the thyroid gland [3], the parapharyngeal space and skull base [4], the oral cavity in the setting of Aicardi syndrome [8], as well as a series of two boys aged 1 year 7 months and 3 years 5 months with head and neck disease [2,6]. Our patient, aged 5 years, lies at the upper end of the age range usually reported for this location, although later-onset cases, such as a 10-year-old girl with thyroid involvement, have also been described [3].

Diagnostic Pitfall on Imaging

CT and MRI allow characterization of the tumor's origin, locoregional extent, and vascular relationships, but their appearance is not specific. In our case, the highly infiltrative nature of the mass, its heterogeneous appearance, and its massive extension into the deep spaces of the face and neck naturally led to an MRI impression favoring a sarcomatous tumor, a hypothesis made all the more plausible in children given that rhabdomyosarcoma is the most common soft-tissue sarcoma at this age and in this anatomical region. This observation illustrates the limits of cross-sectional imaging in differentiating yolk sac tumor from head and neck sarcoma, and underscores the systematic need for biological and histological confirmation before any definitive diagnostic conclusion.

Value of Alpha-Fetoprotein Measurement

AFP is physiologically synthesized by the fetal yolk sac, liver, and gastrointestinal tract [10]. Its production by yolk sac tumor cells makes it a highly valuable diagnostic marker, also used for treatment monitoring and recurrence detection [10]. In our case, the AFP level was markedly elevated (31,087 IU/l), while beta-HCG remained normal, suggesting a pure yolk sac component and allowing choriocarcinomatous differentiation to be excluded. The main biological differential diagnosis remains hepatoblastoma, another pediatric tumor associated with markedly elevated AFP; clinical context, anatomical location, and histology readily distinguish between the two. In any pediatric head and neck mass with an aggressive course, particularly when imaging suggests a sarcoma, AFP measurement deserves to be systematically included in the first-line work-up alongside biopsy: simple, inexpensive, and widely available, it can strongly orient the diagnosis even before histopathological results are available.

Histology and Immunohistochemistry

Yolk sac tumor is histologically characterized by considerable architectural diversity (microcystic/reticular, papillary, solid, polyvesicular vitelline, endodermal sinus patterns), with Schiller-Duval bodies representing its most specific histological hallmark, as observed in our case. The typical immunohistochemical profile combines positivity for AFP and SALL4, focal or membranous positivity for PLAP, and negativity for OCT3/4 and CD30, allowing exclusion of seminoma/germinoma and embryonal carcinoma, respectively, a profile fully consistent with that observed in our patient, confirming the diagnosis of pure yolk sac tumor.

Management and Prognosis

Standard management of yolk sac tumors combines, whenever feasible, surgical resection with platinum-based chemotherapy (classically cisplatin, etoposide, and bleomycin), a strategy that has markedly improved the prognosis of localized gonadal forms, with cure rates exceeding 80% [11]. The prognosis of extragonadal head and neck forms appears more guarded, owing to the difficulty of achieving complete resection given the complex anatomical relationships of this region, and to the risk of delayed diagnosis related to the rarity of this entity. Our patient's course, marked by death within a few days before chemotherapy could even be initiated, illustrates the potentially fulminant nature of the disease when diagnosed at a very advanced locoregional stage, and underscores the importance of the shortest possible multidisciplinary care pathway (otolaryngology – radiology – pathology – pediatric oncology) from the moment of clinical suspicion.

Differential Diagnosis

In a rapidly progressive, infiltrative pediatric head and neck mass, the main differential diagnoses to consider include rhabdomyosarcoma, lymphoma, metastatic neuroblastoma, and other germ cell tumor subtypes (embryonal carcinoma, choriocarcinoma, immature teratoma, or teratoma with a yolk sac component). Combining tumor marker measurement (AFP, beta-HCG, LDH) with biopsy and immunohistochemistry allows, in the vast majority of cases, differentiation between these entities.

Conclusion

Extragonadal head and neck yolk sac tumor is an exceptional entity in children, whose clinical and radiological presentation can mimic a soft-tissue sarcoma, exposing patients to a potentially harmful diagnostic delay given the sometimes fulminant course of the disease. Alpha-fetoprotein measurement, simple and inexpensive, should be systematically combined with biopsy in any pediatric head and neck mass showing rapid growth and an aggressive imaging pattern, in order not to miss this diagnosis and to allow the earliest possible oncologic referral.

Declarations

Conflicts of interest: the authors declare no conflict of interest related to this article.

Funding: this work received no specific funding.

References

1. Makungu H, Ndunguru M, Kwikima U, R. Lyimo F, Scanlan T, et al. Orbital Yolk Sac Tumor in a 2-Year-Old Female Child Presenting with Proptosis—A Rare Entity. Case Rep Radiol. 2026. 2026: 1490793.

2. Roy M, Agarwal S, Gupta A, Bakhshi S, Seith Bhalla A, et al. Extragonadal yolk sac tumor of the head and neck region: a report of two cases. J Cancer Res Ther. 2015. 11: 1000-1002.
3. Furtado LV, Leventaki V, Layfield LJ, Lowichik A, Muntz HR, Pysher TJ. Yolk sac tumor of the thyroid gland: a case report. Pediatr Dev Pathol. 2011. 14: 475-479.
4. Hauser LJ, Chiang T, Ramakrishnan VR, Lovell MA, Kelley PE. Parapharyngeal and skull base yolk sac tumor: a case report with lessons in diagnosis and management. Int J Pediatr Otorhinolaryngol. 2014. 78: 2003-2006.
5. Stang A, Trabert B, Wentzensen N, Cook MB, Rusner C, et al. Gonadal and extragonadal germ cell tumors in the United States, 1973-2007. Int J Androl. 2012. 35: 616-625.
6. Kusumakumari P, Geetha N, Chellam VG, Nair MK. Endodermal sinus tumors in the head and neck region. Med Pediatr Oncol. 1997. 29: 303-307.
7. Bernbeck B, Schneider DT, Koch S, Teske C, Lentrodt J, et al. MAKEI-Study Group. Germ cell tumors of the head and neck: report from the MAKEI Study Group. Pediatr Blood Cancer. 2009. 52: 223-226.
8. Burch-Smith R, Ordonez NG, Ginsberg LE, Ater JL, El Naggar AK. Oral extragonadal yolk sac tumor in a patient with Aicardi syndrome: putative origin and differential diagnosis. Hum Pathol. 2012. 43: 939-942.
9. Epperson MV, Born HL, Wang D, Myer CM. 4th. Yolk Sac Tumors of the Head and Neck in Aicardi Syndrome. Ann Otol Rhinol Laryngol. 2020. 129: 301-305.
10. Canadian Cancer Society. Alpha-fetoprotein (AFP) tumor marker test. [Online resource, accessed 2026].