Tumours that originate in the ductal or secretory epithelial cells of salivary gland are exceedingly rare in children < 2 years of age.\(^1\) A group of tumours has been recognized which usually presents at birth or shortly thereafter, and are composed of basaloid and myoepithelial cells that recapitulate the developing salivary anlage. These tumours have been reported under a variety of names such as congenital basal cell adenoma, basal cell adenoma, basaloid adenocarcinoma, and congenital hybrid basal cell adenoma-adenoid cystic carcinoma, etc.\(^2\) Vawter and Tefft\(^3\) suggested the term embryoma in 1966 for this unique perinatal tumour. Alternatively in 1988, Taylor\(^4\) suggested the term sialoblastoma, because it conveyed both the dysontogenetic character as well as the tumour site within a single name. In extensive PubMed search with keywords “sialoblastoma, embryoma, congenital basal adenoma”, we got 46 cases of “sialoblastoma/embryoma/congenital basal adenoma”, with one case was of German literature and three additional cases of adult sialoblastoma. This article has extensively reviewed the clinical, histopathological and immunohistochemical features, Magnetic resonance imaging (MRI) and Computerized Tomography (CT) findings, treatment and prognosis.

**INTRODUCTION**

Sialoblastoma is a rare congenital tumour of the salivary glands arising mainly from the parotid gland. It is usually diagnosed at birth or shortly thereafter with a significant variability in histological appearance and clinical course. In extensive search of PubMed indexed journals, we got 46 cases of “sialoblastoma/embryoma/congenital basal adenoma”, with one case was of German literature and three additional cases of adult sialoblastoma. This article has extensively reviewed the clinical, histopathological and immunohistochemical features, Magnetic resonance imaging (MRI) and Computerized Tomography (CT) findings, treatment and prognosis.

**DISCUSSION**

**Epidemiology**

Most tumours have been diagnosed at birth; cases from 34 week of fetus to four year of age have been identified in English language literature. (Tables 1 and 2) A Case of 7 year/F reported in German literature.\(^7\) 29 cases (63.03%) were of less than 10 days of age. Male 22, female 23, almost equally affected. In one case sex has not been mentioned. 31 tumour were related to parotid,\(^1,7-29\) 11 submandibular,\(^12,18,30-36\) two cheek minor salivary gland,\(^3,37\) one was in relation with eyelid minor salivary gland\(^38\) and one presented as face and neck mass\(^39\). In addition three cases of adult sialoblastoma were identified. We have reviewed 40 cases tabulated by Saffari et al.\(^5\) in 2011 [Table 1]. Additional 6 cases we have reviewed 46 cases of sialoblastoma/embryoma/congenital basal cell adenoma [Table 2]. 3 adult sialoblastoma\(^6\) cases were also reviewed [Table 2].

**Clinical features**

Clinically most of babies presented as cheek and submandibular mass. The reported size of tumor ranges from a peanut to a maximum of 15 cm in diameter.\(^28,39\) At one occasion it was associated with superficial haemorrhage and necrosis.\(^39\) The clinical
Early conservative surgical approach

Two further recurrence following 30 mo

CT appearance is of a soft-tissue mass hypodense to the brain and isodense to muscle. Som et al.\textsuperscript{14} detected low–intermediate signal intensity and slightly higher intermediate signal intensity...
on T1-weighted (T1-W) and T2-weighted (T2-W) images, respectively. In a case reported by Yekeler et al., the greater part of the lesion excluding necrotic and haemorrhagic areas was mildly hyper intense on T2-W images, which was lower than that described by Som et al. The finding of mild hyperintensity on T2-W images suggests a high nucleus/cytoplasmic ratio belonging to blastoma and can be predictive for the diagnosis of blastomas. In their case, the cause of intralesional haemorrhage on MRI was unclear. It could have occurred spontaneously into the fragile tumour tissue or have resulted from minor trauma during vaginal delivery.

**Histopathological features**

The morphology of sialoblastoma is very characteristic with the presence of nests of basaloid cells with a palisading pattern at the periphery and maturation toward the centre. Sialoblastomas may have admixed histologic appearance, ranging from benign hamartomatous lesions with marked similarity to normal fetal salivary gland tissue to highly malignant tumour. Batsakis and colleague proposed histologic criteria for assessment of malignancy in a sialoblastoma, which included “invasion of nerves or vascular spaces and ancillary findings of necrosis and cytological atypia beyond that expected or presumed for an embryonic epithelium. Mitotic figures are variable, 3-4/High Power Field (HPF), and 6-7/10 HPF to 20/10 HPF has been reported. Areas of necrosis also increased in recurrent cases with focal calcification in isolated cases and necrosis with total tumour replacement of tumour stroma after chemotherapy. Proliferative index from 3cell/10 HPF to 94 cell/10 HPF has been reported.

**Table 2: Cases of paediatric and adult sialoblastoma 2010-2011, 2010* adult SB**

| Year | References | Age/sex | Site | Size (cm) | Treatment | Recurrence/Rx | Metastasis | Histological diagnosis | Follow up DF (Disease free) |
|------|------------|---------|------|-----------|-----------|---------------|------------|------------------------|----------------------------|
| 2011 | [5]        | AT Birth, R cheek | 2 x 2 cm, within 5 days size increase up to 4.5 x 4.5 cm | Surgical excision | No recurrence | No | SB | 7 Month DF |
| 2011 | [29]       | 3 Mo FM L parotid | Not mentioned | 3.5 year age chemo | After 1.5 year, orbital recurrence, total resection, adj radio | Age of 6.5, At 6.5 years of age, metastasis to the right lung. Metastasis to the left lung two years later, 6 months later, third pulmonary recurrence in the right upper lobe | SB | 7 year DF |
| 2010 | [28]       | 18 Mo/FM L parotid | Size increase from peanut to table tennis ball in 1 month (3 x 3 x 3), H/O swelling since birth, operated two months before, noBx Report, swelling gradually increasing in size | Surgical excision | Aft 6 month, 125 I seed implant brachythrapy, complete response | Radical surgery, with chemo, recurrence time not mentioned | No | SB | 21 month DF |
| 2010 | [26]       | 12 Mo/M R cheek | Surgical excision | FNA diagnosis was PA, spontaneous regression after FNA, 4 year later 3 cm mass | Surgical excision | 1 year later recurred (6 x 7 x 7) | Rt and Lt lung met, chemo, complete response | SB | 12 months DF |
| 2010 | [27]       | At Birth/F L Parotid | Surgical excision | FNA diagnosis was PA, spontaneous regression after FNA, 4 year later 3 cm mass | Surgical excision | 1 year later (6 x 7 x 7) | SB | 12 months DF |
| 2010 | [39]       | 3 days/M L face and neck mass | 15 x 10 x 8 (superficial haemorrhage necrosis) | Sx, margin +ve | Recurrence after 3 month, treated with chemotherapy | No | No | 4 year |
| 2010* | [6] | 46 year/FM R Parotid | 3 x 2 cm six month duration | Sx | 15 year later lump in surgical scar, after 5 treated with Sx and radio Incision biopsy | Tumor did not progress till last 8 month, surgery avoided due to patient factors | No | 7 year DF |
| 2 | 83/M Palate | 2.5 x 2.5 cm, exact duration did not mentioned | Sx | Recurrence after 3 month, treated with chemotherapy | No | No | No | No follow up detail |
| 3 | 55/M Palate | No clinical data | No | No | No | No | No | No |

*Year of publication, FT: Free of tumour, NA: Not available, Ref: Reference, M: Male, F: Female, R: Right, L: Left, w: Week, mo: Month, Sx: Surgical excision, DF: Disease free, SB: Sialoblastoma
gland; 2 lesions were from the submandibular gland. All lesions presented as nodular to multinodular swellings and ranged in size from 2.0 to 7.0 cm. The principal sign or symptom was rapid growth. Two histologic patterns with differing behaviour predominated: (1) A favourable pattern had semi encapsulation of cytologically benign basaloid tumour cells with intervening stroma; and (2) an unfavourable histology of anaplastic basaloid tumour cells, minimal stroma, and broad pushing to infiltrative periphery. Four and three tumours had favourable and unfavourable growth patterns, respectively. One unfavourable lesion had vascular invasion, and another demonstrated perineural invasion. All three tumours with unfavourable histology recurred. Tumour cells in three cases were immunohistochemically reactive for keratin, S-100, smooth muscle actin, and calponin to varying degrees. All three tumours were reactive for p63. AFP was expressed in two unfavourable tumours. AFP positivity also reported by xx. Ki67 was expressed at 3% in a favourable tumour and 40% and 80% in the two unfavourable lesions. Yasi et al. has reported Ki-67 (30%) in their case. As a prognostic marker, Ki67 immunostaining demonstrated expected findings of a lower proliferative index in the favourable tumour in one case as opposed to significantly higher indices in unfavourable tumours two cases. In the case study by Brandwein et al. Ki67 developed as a significant finding in the course of the recurrences of tumour. The incorporation of Ki67 index with the favourable/unfavourable histologic indicator may be useful for the prognosis of these tumours. So immunohistochemical these tumour express S-100 and vimentin diffusely. Cytokeratin accentuated the ductal structure.

**Recurrence and metastasis**

10 cases recurred after initial treatment. Most of them recurred within 24 month. Four cases of lung metastasis with two cases have multiple lung metastasis and two cases of cervical lymph node metastasis have been reported. In reported cases no case succumbed to death due to recurrence or metastasis [Tables 1 and 2].

**Treatment**

Various treatment modalities like surgical excision, chemotherapy, radiotherapy and combination for primaries, recurrent and metastatic cases have been mentioned. One case spontaneously regresses after FNAC till one year than increased in size, treated with superficial parotidectomy and recurred. After metastasis to lung treated with chemotherapy and got complete response. One case treated with 125I brachytherapy cured completely [Table 1 and 2].

**Prognosis**

Only three cases succumbed to death; due to septicaemia and unrelated to sialoblastoma, one in each category. In reported literature maximum of 43 year of disease free survival has been reported. In most of cases functional and aesthetic efficacy has been achieved.

**Cytogenetics**

Ozdemir et al. reported a case of congenital sialoblastoma presenting with the PCD and a high level of AFP, which associations have not previously been reported.

**Adult sialoblastoma**

Essentially, sialoblastoma is a disease of infancy with the oldest case presenting at four and seven year of age in English and German literature respectively [Tables 1 and 2]. About one third of pediatric sialoblastoma cases will have a cribriform growth pattern. No adult cases have been reported with a specific diagnosis of sialoblastoma. If even focal cribriforming were present, such cases have undoubtedly been diagnosed as adenoid cystic carcinoma. Such was the circumstance in the three adult tumours presented by Dardic et al. Each case, however, has the primitive histopathology with discrete nests of basaloid tumour cells, associated bilayered ductal structures and the fibromyxoid stroma characteristic for sialoblastoma with its resemblance to fetal salivary gland or salivary gland with arrested development. Sialoblastoma, whether in a child or adult with or without a cribriform growth pattern, appears to have a more favourable prognosis than adenoid cystic carcinoma. Detailed data mentioned in Table 1 (2010 * 1, 2 and 3).

In adult sialoblastoma cases, cribriform histology needs to be tempered with the overall primitive organization of the basaloid tumor cells, often with associated single or branching ductal structures, enclosed by loose collagenous to myxoid stroma. The latter aspects produce a likeness to developing salivary gland in the fetus and, as a hallmark of sialoblastoma, are an essential diagnostic feature. It is notable that sialoblastomas are generally circumscribed and even partially encapsulated. A cribriform growth pattern does not necessarily imply adenoid cystic carcinoma with its inherently poor long-term prognosis. The synthesis of excess glycosaminoglycans myoepithelial cells - responsible for discrete intercellular spaces or histologic variants associated with this process-is common to a number of salivary gland tumors, including pleomorphic adenoma, myoepithelioma, basal cell adenoma, epithelial-myoepithelial carcinoma, and polymorphous low-grade adenocarcinoma. Ultrastructural studies of some cases of sialoblastoma reveal reduplication of basal lamina, and in one adult example in this report, frank intercellular accumulations of glycosaminoglycans and basal lamina in association with basaloid tumor cells.
and basal lamina by neoplastic basal/Sialoblastoma is, therefore, another salivary gland tumor with a potential for a cribriform element.

**Conclusion**

Sialoblastoma is rare salivary gland tumours, almost all cases have been reported below four year of age in English literature. Recently cases of adult sialoblastoma have been reported. And it has been suggested that cribriform pattern is evident in sialoblastoma. Surgical excision with negative margin is mainstay of treatment, yet in unresectable case brachytherapy has been used successfully. Instead of rapid growth potential prognosis is good; no death has been reported due to metastasis. Existence of adult sialoblastoma needed further clarification with large case series.

**References**

1. Batsakis JG, Frankenthaler R. Embryoma (sialoblastoma) of salivary glands. Ann Otol Rhinol Laryngol 1992;101:958-60.
2. Leon B, John WE, Peter R, David S. Pathology and genetic. WHO Head and Neck Tumor. 5th ed. Brandwein Ginsler. Sialoblastoma. 2005. p. 253.
3. Vawter GF, Tefft M. Congenital tumors of the parotid gland. Arch Pathol 1966;82:242-5.
4. Taylor GP. Congenital epithelial tumor of the parotid-sialoblastoma. Pediatr Pathol 1988:8:447-52.
5. Saffari Y, Blei F, Warren SM, Milla S, Greco MA. Congenital minor salivary gland sialoblastoma: A case report and review of the literature. Fetal Pediatr Pathol 2011;30:32-9.
6. Dardick I, Thomas TD, McComb RJ. Sialoblastoma in adults: Distinction from adenoid cystic Carcinoma. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 2010;109:109-16.
7. Seifert G, Donath K. Juvenile pleomorphic parotid adenoma of embryonal structure. Pathol 1998;19:286-91.
8. Tatitlede S, Karsidag S, Ugurlu K, Sadikoglu B, Tanik C, Bas L. Sialoblastoma: A congenital epithelial tumor of the salivary gland. J Pediatr Surg 2006;41:1322-5.
9. Yekeler E, Dursun M, Gun F, Kilincaslan H, Ucar A, Gencelac H, et al. Sialoblastoma: MRI findings. Pediatr Radiol 2004;34:1005-7.
10. Huang R, Jaffer S. Imprint cytology of metastatic sialoblastoma. A case report. Acta Cytol 2003;47:1123-6.
11. Siddiqi SH, Solomon MP, Haller JO. Sialoblastoma and hepatoblastoma in a neonate. Pediatr Radiol 2000;30:349-51.
12. Seifert G, Donath K. The congenital basal adenoma of salivary glands. Contribution to the differential diagnosis of congenital salivary gland tumours. Virchows Arch 1997;430:311-9.
13. Brandwein M, Al-Naeef NS, Manwani D, Som P, Goldfelder L, Rothschild M, et al. Sialoblastoma: Clinicopathological/immunohistochemical study. Am J Surg Pathol 1993;23:324-48.
14. Som PM, Brandwein M, Silvers AR Rothschild MA. Sialoblastoma (embryoma): MR findings of a rare pediatric salivary gland tumor. AJNR Am J Neuroradiol 1997;18:487-50.
15. Alvarez-Mendoza A, Calderon-Elvir C, Carrasco-Daza D. Diagnostic and therapeutic approach to sialoblastoma: Report of a case. J Pediatr Surg 1999;34:1875-7.
16. Hsueh C, Gonzalez-Crussi F. Sialoblastoma: A case report and review of the literature on congenital epithelial tumors of salivary gland origin. Pediatr Pathol 1992;12:205-14.
17. Batsakis JG, Mackay B, Ryka AF, Seifert RW. Perinatal salivary gland tumours (embryomas). J Laryngol Otol 1988;102:1007-11.
18. Williams SB, Ellis GL, Warnock GR. Sialoblastoma: A clinicopathologic and immunohistochemical study of 7 cases. Ann Diagn Pathol 2006;10:320-6.
19. Solomon MR, Kaleem Z, Chen CK. Concurrent sialoblastoma (embryoma) of parotid gland and hepatoblastoma (fetal type) arising in a new born: A previously unreported association (abstract). Congress of the International Association of Oral Pathologists (IAOP), York England, July 1994, Abstract P37.
20. Casas LA, Gonzalez-Crussi F, Pensler JM. Monomorphic adenoma of the parotid in a premature neonate. Ann Plast Surg 1989;22:47-9.
21. Roth A, Michean C. Embryoma (or embryonal tumor) of the parotid gland: Report of two cases. Pediatr Pathol 1986;5:9.
22. Canalis RF, Mok MW, Fishman SM, Hemenway WG. Congenital basal adenoma of the submandibular gland. Arch Otolaryngol 1980;106:284-6.
23. Stones DK, Jansen JC, Griessler D. Sialoblastoma and hepatoblastoma in a new born infant. Pediatr Blood Cancer 2008;52:883-5.
24. Scott JX, Krishnan S, Bourne AJ, Williams MP, Agzarian M, Revess T. Treatment of metastatic sialoblastoma with chemotherapy and surgery. Pediatr Blood Cancer 2008;50:134-7.
25. Ozdemir I, Simsek E, Silan F, Demirci F. Congenital sialoblastoma (embryoma) associated with premature centromere division and high level of alpha-fetoprotein. Prenat Diagn 2005;25:687-9.
26. Kattoor J, Baisakh MR, Mathew A, Somanathan T, Nayak N, Abraham EK. Sialoblastoma: A rare salivary gland neoplasm. Indian J Cancer 2010;47:219-20.
27. Prigent M, Teissier N, Peuchmaur M, Maleh-Berges ME, Philippe-Chomette P, Cardin P, et al. Sialoblastoma of salivary glands in children: Chemotherapy should be discussed as an alternative to mutilating surgery. Int J Pediatr Otorhinolaryngol 2010;74:942-5.
28. Shan XF, Cai ZG, Zhang JG, Zhang J, Gao Y, Yu GY. Management of sialoblastoma with surgery and brachytherapy. Pediatr Blood Cancer 2010;55:1427-30.
29. Farooqi KM, Kessel R, Brandwein-Gensler M, Granowetter L, Manwani D. Sialoblastoma-long-term follow-up and remission for a rare salivary malignancy. Rare Tumors 2011;3:e13.
30. Harris MD, McKeevcr P, Rohcrstson JM. Congenital tumours of the salivary gland: A case report and review. Histopathology 1996;17:155-7.
31. Vidyadhar M, Amaand C, Thuan Q, Prabhakaran K, Sialoblastoma. J Pediatr Surg 2008;43:e11-3.
32. Cristofaro M, Giudice A, Amenta M, Giudice M. Diagnostic and therapeutic approach to sialoblastoma of submandibular gland: A case report. J Oral Maxillofac Surg 2008;66:123-6.
33. Verret DJ, Galindo RL, DeFatta RJ, Bauer PW. Sialoblastoma: A rare submandibular gland neoplasm. Ear Nose Throat J 2006;85:440-2.
34. Garrido A, Humphrey G, Squire RS, Nishikawa H. Sialoblastoma. Br J Plast Surg 2000;53:697-9.
35. Green RS, Tunkel DE, Small D, Westra WH, Argani P. Sialoblastoma: Association with cutaneous hamartom (organoid nevus)? Pediatr Dev Pathol 2003;6:504-5.
36. Marucci DD, Lawson K, Harper J, Sebire NJ, Dunaway DJ. Sialoblastoma: A rare sialoblastoma-like tumor with a sarcomatoid myoepithelial component. Oral Oncol 2009;62:e241-60.
37. Stones DK, Jansen JC, Griessler D. Sialoblastoma and hepatoblastoma in a new born infant. Pediatr Blood Cancer 2008;50:134-7.
38. Shet T, Ramadwar M, Sharma S, Laskar S, Arora B, Kurkure P. An eyelid sialoblastoma-like tumor with a sarcomatoid myoepithelial component. Pediatr Dev Pathol 2007;10:309-14.
39. Saribeyoglu ET, Devecioğlu O, Karakas Z, Anak S, Unuvar A, Ağaoglu L, et al. How to manage an unresectable or recurrent sialoblastoma. Pediatr Blood Cancer 2010;55:374-6.
40. Adinolfi A, Adinolfi M, Lessof. Alpha-feto-protein during development and in disease. J Med Genet 1975;12:138-51.
41. Mann JR, Raafat F, Robinson K, Imeson J, Gornall P, Sokal M, et al. The United Kingdom children’s cancer study group’s second germ cell tumor study: Carboplatin, etoposide, and bleomycin are effective treatment for children with malignant extracranial germ cell tumors, with acceptable toxicity. J Clin Oncol 2000;18:3809-18.

42. Pitkanen S, Salo MK, Kuusela P, Holmberg C, Simell O, Heikinheimo M. Serum levels of oncofetal markers CA 125, CA 19-9, and alpha-fetoprotein in children with hereditary tyrosinemia type I. Pediatr Res 1994;35:205-8.

43. Dardick I. Color atlas/text of salivary gland tumor pathology. vol. 83. New York: Igaku-Shoin Medical Publishers, Inc; 1996. p. 215.

44. Grenco RT, Abendroth CS, Davis AT, Levin RJ, Dardick I. Hybrid tumors or salivary gland tumors sharing a common pathway? Reexamining adenoid cystic and epithelial-myoepithelial carcinomas. Oral Surg Oral Med Oral Pathol Oral Radiol Endod 1998;86:188-95.

45. Boór A, Jurkovic I, Kocan P, Jenca A. Collagenous spherulosis in epithelial-myoepithelial carcinoma of the parotid gland: Histological and immunohistochemical study of a case. ORL J Otorhinolaryngol Relat Spec 2002;64:148-51.

46. Araújo VC, Loducca SV, Sousa SO, Williams DM, Araújo NS. The cribriform features of adenoid cystic carcinoma and polymorphous low-grade adenocarcinoma: Cytokeratin and integrin expression. Ann Diagn Pathol 2001;5:330-4.

47. Simpson PR, Rutledge JC, Schaefer SD, Anderson RC. Congenital hybrid basal cell adenoma-adenoid cystic carcinoma of the salivary gland. Pediatr Pathol 1986;6:199-208.

48. Daley TD, Dardick I. An unusual parotid tumor with histogenetic implications for salivary gland neoplasms. Oral Surg Oral Med Oral Pathol 1983;55:374-81.

49. Ortiz-Hidalgo C, de León-Bojorge B, Fernandez-Sobrero G, Sánchez Marle JI, Martin del Campo N. Sialoblastoma: Report of a congenital case with dysembryogenic alterations of adjacent parotid gland. Histopathology 2001;38:79-80.

50. Krolls SO, Trodahl JN, Boyers RC. Salivary gland lesions in children. A survey of 430 cases. Cancer 1972;30:459-69.

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