

A RARE SALIVARY GLAND TUMOUR OF THE NASAL SEPTUM:AN ACINIC CELL CARCINOMA

Type of article- Case report

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Abstract-

AIM- To report a rare case of salivary gland tumour of the nasal cavity originating from the nasal septum.

BACKGROUND -Acinic cell carcinoma of the nasal cavity is an exceptionally rare tumour. These tumors account for almost about 1% of all salivary gland neoplasms .It also accounts for about 10-15% of all malignant salivary gland neoplasms.

CASE DESCRIPTION- We present a case of 42-year-old female patient who presented with complaints of left sided nasal bleeding and nasal obstruction. On further evaluation, she was diagnosed with rare salivary gland tumour, an Acinic cell carcinoma originating in the nasal septum of the left nasal cavity. She underwent left lateral rhinotomy with complete resection of tumour and regular follow-up with no further radiotherapy intervention.

CONCLUSION - Acinic cell carcinomas are very slowly progressive, low grade malignant tumour with good prognosis. However, surgery remains the main stay of treatment and radiotherapy post-operatively to prevent micro metastasis and recurrence. In our case surgery was the only modality of treatment for this tumor in this case as there is no recurrence of tumor till date.

Keywords- Acinic cell carcinoma, Epistaxis, Nasal septum, Malignant, Lateral rhinotomy

INTRODUCTION

Acinic cell carcinoma is a uncommon type of salivary gland tumour which comprises of 6-8% of all salivary malignancies. It is most commonly seen arising from the major salivary glands like parotid gland. It sometimes also arise in the minor salivary glands which includes lips, palate, buccal mucosa and other ectopic sites^[1]. Most common site in minor salivary glands being the oral cavity^[2]. Salivary gland neoplasms are not commonly seen arising in the nasal cavity. The most common type of benign salivary gland tumor in the nasal cavity is pleomorphic adenoma and malignant is adenoid cystic carcinoma. Primary nasal acinic cell carcinoma is rare, accounting for only 1% to 4% of all malignant nasal neoplasms^[3]. According to the World Health Organisation acinic cell carcinoma is a malignant epithelial neoplasm of the salivary glands, in which few neoplastic cells demonstrate serous acinar cell differentiation characterized by cytoplasmic zymogen secretory granules^[4]. Histologically the tumor is characterized by basophilic granular cells arranged in acinic pattern similar to normal acinar tissue of parotid gland^[5]. It is less aggressive with best survival rate of any other salivary malignancy. Cases of recurrence and metastasis in regional as well as in distant sites like bone, lung and brain are reported^[6]. Here we present a case of rare salivary gland tumour a primary nasal acinic cell carcinoma

CASE REPORT

A 42 year old female patient who presented to ENT department in tertiary health care centre on 13 september 2020, with complaints of obstruction in the left side nasal cavity and intermittent episodes of nasal bleed left side since 1 year. It was associated with left sided nasal discharge and headache from 6 months. She had similar complaints 5 years back no mass was seen on clinical examination. On radiological examination showed deviated nasal septum with chronic sinusitis features for which she underwent septoplasty with functional endoscopic sinus surgery and remained asymptomatic till 4 years. Now on examination of the external framework showed widening over the left side of the dorsum with intercanthal distance being within normal limits (Figure 1). Anterior Rhinoscopic examination revealed single pinkish fleshy mass with irregular surface covered with discharge and slough filling entire nasal cavity extending anteriorly upto nasal vestibule (Figure 2). On probing mass is insensitive, firm in consistency, bleeds on touch, not friable, able to pass all around except laterally. Examination of the right nasal cavity showed septum deviated to right side, floor and turbinates were normal. Posterior Rhinoscopy, Para Nasal Sinus and face examination were within normal limits. Examination of Ear, Throat, neck and other systems was unremarkable. Diagnostic nasal endoscopy was done and the clinical findings were confirmed. All the routine biochemical parameters were within normal range.

A biopsy was taken from the nasal mass and the specimen was sent for histopathology, which revealed tissue containing areas of necrosis and tumour cells organised in nests, trabeculae and glandular pattern. It has small round nucleus and moderate bluish cytoplasm, suggestive of features of acinic cell carcinoma (FIGURE 3).

A Computed tomography scan of nose and paranasal sinuses showed a heterogeneously enhancing soft tissue density lesion noted epicentered in the left nasal cavity measuring 4.8 x 1.8 x 3 cm extending anteriorly upto ala of nose, posteriorly upto left nasal choana, superiorly infiltrating tip of middle turbinate, medially infiltrating the anterior part of nasal septum with erosion of bone, laterally infiltrating and causing erosion of left inferior turbinate causing mass effect on medial wall of maxillary sinus left

side causing bulging and thinning of wall. It also shows erosion of palatine process of maxilla of left side inferiorly. There is no cervical lymph node involvement. The features were suggestive of a malignant neoplastic lesion (FIGURE 4&5). According to American Joint Committee on Cancer (AJCC) TNM classification of Malignant Tumours of nasal cavity the patient was staged as T2 N0 Mx [7].

Patient under general anaesthesia underwent excision of the nasal mass with left lateral rhinotomy approach (FIGURE 6). Intraoperatively endoscopy showed that the mass was seen arising from the septal mucosa involving entire left side nasal cavity extending posterior upto choana and involving septum causing erosion of septal bone and cartilage and extending to opposite side (FIGURE 7). The tumour was excised in toto along with the area involving cartilaginous part of septum, maxillary crest and perpendicular plate of ethmoid (FIGURE 8). Specimen was sent for histopathological examination. Prophylactically no treatment was performed for neck as there was no cervical lymph nodes involvement both clinically and radiologically. The post-operative course was uneventful.

Histopathological examination showed pleomorphic acinar and ductal cells arranged in follicular pattern, solid sheets, microcystic, macrocystic spaces with papillary proliferation. With Periodic Acid Schiff staining cells were large polygonal having basophilic granular cytoplasm with round eccentric nuclei features suggesting acinic cell carcinoma high grade microcystic type. The margins of the tumour specimen was found to be tumor free (FIGURE 9).

In this case, after surgery no further treatment was carried out. A monthly followup was done upto 6 months diagnostic nasal endoscopic examination was normal. At 6th month diagnostic nasal endoscopy was done which showed granulation tissue over nasal septum for which biopsy was done and sent for histopathological examination which revealed inflammatory granulation tissue with no evidence of recurrence. A followup CT scan was performed after one year of surgery which showed no evidence of local recurrence or metastasis. The patient has been followed-up for almost every 6 months till date and there is no complaints of any symptoms, local recurrence or regional metastasis has been encountered so far.

DISCUSSION

Malignant neoplasms of the nose and paranasal sinuses are considered to be rare. It accounts for about 3% of all head and neck tumours [8]. Acinic cell carcinoma is an exceptional, slow-growing, less aggressive malignant tumor of the salivary gland, primarily the parotid gland [9]. It affects only 0.3-0.5% of all the neoplasms of minor salivary glands. It is most commonly localised in the oral cavity. The nasal acinic cell carcinoma is extremely rare. There are few cases reported previously in the literature. The cases reported so far details are outlined below (TABLE 1).

STUDIES	AGE	SEX	SITE OF ORIGIN	SYMPTOMS	TREATMENT (Surgery, Radiotherapy)	FOLLOW UP
Present study	42	F	Nasal septum	Nasal obstruction, epistaxis	Surgery	1.5 years
Dionísio S et al, 2021 [10]	62	M	Nasal lateral wall	Nasal obstruction, epistaxis	Both	6 months
Naseeruddin et al, 2019 [11]	84	M	Nasal septum	Nasal obstruction, epistaxis	Both	1 year
Hou et al., 2017 [12]	62	M	Nasal lateral wall	Nasal obstruction, epistaxis and hyposmia	Both	2 years
P S Jayalakshmy et al, 2016 [13]	52	M	Middle turbinate	Nasal obstruction, epistaxis	Surgery	N/S
Gangadhara Somayaji et al, 2014 [14]	65	F	Nasal lateral wall	Nasal obstruction, epistaxis	Surgery	1 year
D Ravi et al, 2012 [15]	50	F	Inferior turbinate	Nasal obstruction	Surgery	4 years
Hammami et al., 2010 [16]	47	F	Nasal septum	Nasal obstruction, Hyposmia	Both	4 years
Manganaris et al., 2010 [17]	51	F	Vestibule	Pain	Surgery	3 Years
Neto et al., 2005 [18]	60	F	Superior meatus	Nasal obstruction	Surgery	17 years
Neto et al., 2005	42	F	Inferior turbinate	Nasal obstruction	Surgery	7 years

[18]						
Neto et al., 2005 [18]	65	M	Inferior turbinate	Nasal obstruction	Surgery	4 years
Neto et al., 2005 [18]	50	M	Nasal cavity	Nasal obstruction	Both	12 years and recurrence
Sapçi et al., 2000 [19]	47	M	Nasal septum	Nasal obstruction,epistaxis	Surgery	1.5 years
Jasin et al., 1999 [20]	44	F	Nasal septum	N/S	Surgery	2.5 years
Von Biberstein et al., 1999 [21]	76	F	Middle turbinate	Nasal mass	Surgery	3 years
Schmitt et al., 1994 [22]	60	M	Inferior turbinate	Nasal obstruction	N/S	N/S
Valerdiz-Casasola et al., 1993 [23]	47	M	Nasal cavity	Nasal obstruction,epistaxis	Both	10 months
Takimoto et al., 1989 [24]	60	F	Middle and inferior turbinate	Nasal mass,epistaxis	Surgery	2 years
Hanada et al., 1988 [25]	68	M	Inferior turbinate	Nasal obstruction	Both	3 years
Finkelhor et al., 1987 [26]	45	F	Nasal septum	Nasal obstruction	Surgery	N/S
Ordenez et al., 1986 [27]	60	F	Superior meatus	Nasal mass,epistaxis	Surgery	7 years
Perzin et al., 1981 [28]	75	F	Inferior turbinate	Nasal obstruction,epistaxis	Surgery	N/S

TABLE 1-Reviewed cases of nasal acinic cell carcinoma

M: male; F: female; N/S: not specified

The review of literature included 23 cases including the present study case. The age wise distribution shows age ranging from 42-84 years and the maximum patients belong to age group between 42-50 years. The studies showed females preponderance (57%). The most common site of origin was found to be the turbinates in 9 cases, 7 cases in the nasal septum, 3 cases in lateral nasal wall, 2 cases in the nasal cavity and only 1 cases were reported each in the vestibule and superior meatus. The most common complaint was nasal obstruction (82%), followed by epistaxis (50%), hyposmia (9%), nasal mass and nasal pain 1 case study. The diagnosis of all cases were based entirely on histopathological examination.

As per the review study done by Ellis and Cori of 294 cases at Armed Forces Institute of Pathology of acinic cell carcinoma showed that the parotid gland was the most common site for tumour origin. The histologic subtypes reported solid (38%), microcystic (33%), papillary cystic (25%) and follicular (4%), which was similar to other studies [29]. Batsakis et al. proposed a three-tiered grading system to histologic variants: Solid-lobular, acinar-microcystic, pilocystic, tubuloductal, follicular and dedifferentiated [30]. Batsakis et al. divided the tumor histologically into high and low-grade lesions. High-grade lesions demonstrate aggressive local invasion, extraparenchymal invasion, medullary growth architecture and prominence of undifferentiated cells. Abrams et al. reported histologic variants as solid, microcystic, papillary-cystic and follicular growth patterns [31]. The cellular features were classified as acinar, intercalated ductal, vacuolated, clear and nonspecific glandular. Our case showed an acinar cell type arranged in sheets and trabeculae.

The main modality of the treatment is complete surgical resection. Post-operative radiation is usually not recommended for low-grade salivary malignancies but used for advanced, high grade tumors, recurrent tumors, tumors with positive surgical excision margins or tumors with extensive perineural and/or lymphovascular invasion. Chemotherapy for acinic cell carcinoma is ineffectual [12]. Neck dissection is not recommended, because the metastatic involvement of cervical lymph nodes are relatively low, with the reported nodal metastasis rates for acinic cell carcinomas of parotid gland being 5-10% [2].

Lymph node metastasis are observed in 10-16% [2]. Local recurrence was detected in 33%, with better control in lower stage tumors. Distant metastasis developed in 12% of the patients and were usually associated with local recurrence. Survival rates were 83%, 76% and 65% for 5, 10 and 15 years respectively after initial therapy.

In present case study surgery was the main treatment modality with no postoperative chemotherapy or radiotherapy. There was no lymph node metastasis or local recurrence till date.

CONCLUSION

Acinic cell carcinoma arising from heterotrophic salivary cell rests in the nose and paranasal sinuses requires high degree of clinical suspicion and histopathological confirmation for the diagnosis. The treatment regimen for Sino nasal acinic cell carcinomas remains empirical and possibly controversial due to insufficient clinical data. This paper discussed the considerations in the evaluation and management of an exceptionally rare malignancy arising in the nasal cavity. Surgery remains the main stay of treatment and radiotherapy post-operatively helps in preventing micro metastasis and recurrences. The case reported here is one of the not so frequently encountered cases. Our case did not undergo radiotherapy. Hence proving that surgery was the only modality of treatment for this tumor in this case as there is no recurrence of tumor till date. The best result is often achieved when each patient is individually managed based on expected local control rates, functional and cosmetic outcome, risk of complications and individual experience.

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FIGURE 1- SHOWING WIDENING OF LEFT DORSUM OF NOSE



FIGURE 2- SHOWING MASS ON LEFT NASAL CAVITY ON ANTERIOR RHINOSCOPIC EXAMINATION

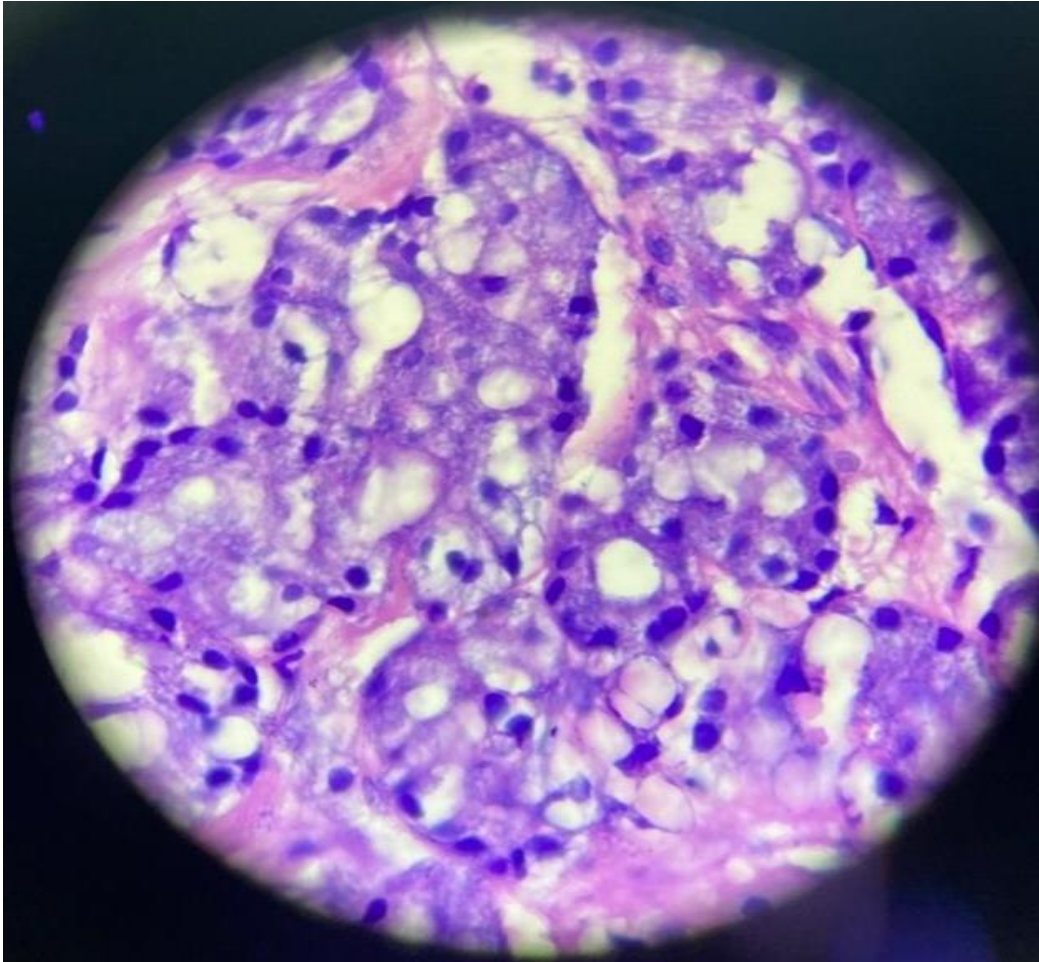


FIGURE 3-SHOWING HISTOPATHOLOGICAL FEATURES OF ACINIC CELL CARCINOMA

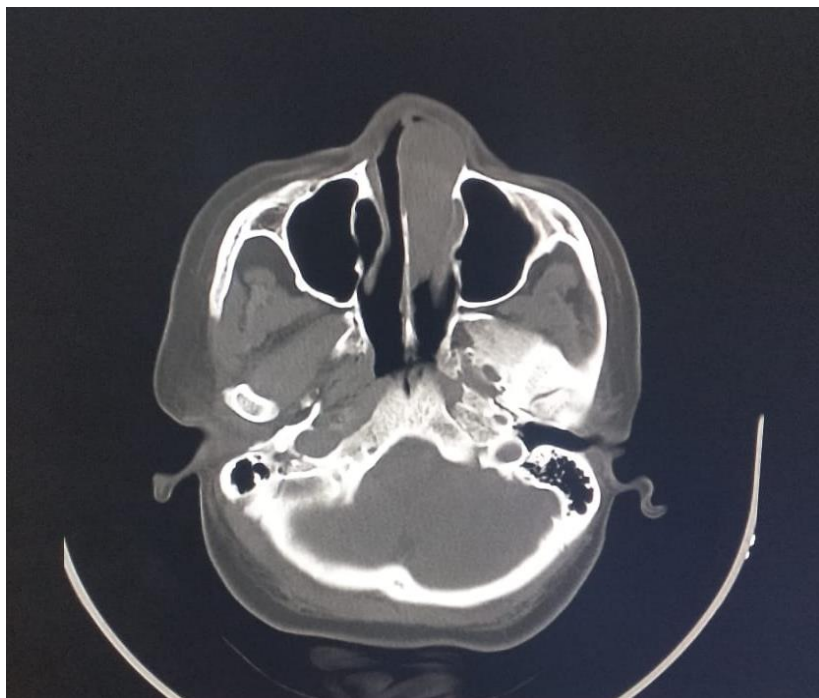


FIGURE 4- COMPUTED TOMOGRAPHY OF NOSE AND PARANASAL SINUS AXIAL VIEW SHOWING MASS IN LEFT SIDE NASAL CAVITY

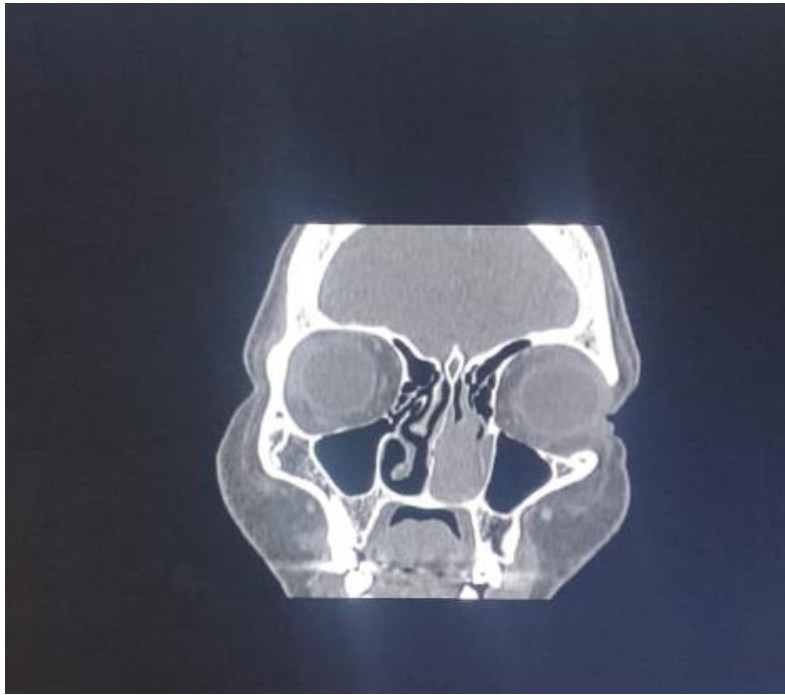


FIGURE 5-COMPUTED TOMOGRAPHY OF NOSE AND PARANASAL SINUS CORONAL VIEW SHOWING MASS IN LEFT SIDE NASAL CAVITY



FIGURE 6-SHOWING LEFT LATERAL RHINOTOMY APPROACH



FIGURE 7-SHOWING DIAGNOSTIC ENDOSCOPY FINDINGS



FIGURE 8- SHOWING EXCISED SPECIMEN

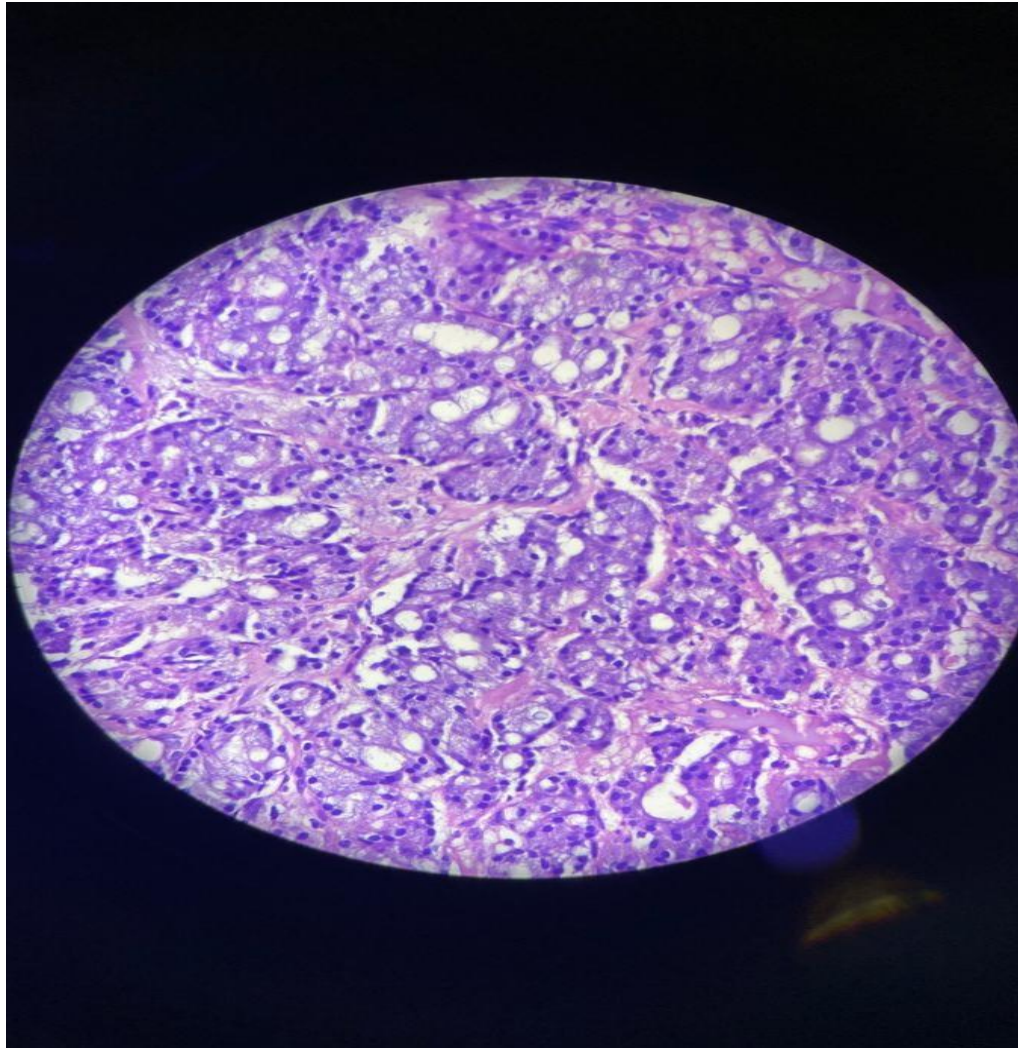


FIGURE 9 SHOWS HISTOPATHOLOGY PICTURE OF ACINIC CELL CARCINOMA WITH BASOPHILIC GRANULAR CYTOPLASM WITH ROUND OR OVAL SHAPED NUCLEI OF TUMOUR CELLS.