Case Report

Insights of Clinical and Medical Images

Rare Endoscopically Resected Sinonasal Adenoid Cystic Carcinoma Masqueraded By Non-Specific Nasal Congestion. A Case Report And Review Of Literature

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C.M: Study conception, revision of the manuscript, proofreading

Highlights:

1. Rare sinonasal adenoid cystic carcinoma
2. Presenting with non-specific features mimicking sinusitis
3. Endoscopic resection to achieve tumour free margins
4. Role of endoscopic evaluation when difficulty to ascertain diagnosis on imaging

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1. Abstract

1.1 Introduction and Importance: Adenoid cystic carcinoma is a head and neck cancer that most often involve the salivary glands however sinonasal adenoid cystic carcinoma is a possible rare entity. They tend to present with nonspecific symptoms hence tend to be treated for recurrent sinusitis before suspicion for neoplasia arises. Mainstay of treatment is surgical resection followed by radiation therapy to minimise risk of reoccurrence.

1.2 Case presentation: 32-year-old female presenting with sinonasal adenoid cystic carcinoma who underwent endoscopic resection and post-operative radiation to achieve tumour free margins without reoccurrence at 2 year follow up. Our experience of endoscopic approach and radiotherapy for diagnosis and curative intent in East Africa
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1.3 Clinical Discussion: We concur with the recommendation to undergo surgical resection followed by radiation for sinonasal adenoid cystic carcinoma. We deployed a Functional endoscopic sinus surgery (FESS) approach whose efficacy of other forms of sinonasal cancers for oncological resection has been comparable to open approach with favourable benefits.

1.4 Conclusion: Sinonasal adenoid cystic carcinoma is a rare carcinoma of the sinonasal region presenting with nonspecific nasal symptoms. We hope through our successful experience of achieving adequate resection via an endoscopic approach followed by radiotherapy to prevent reoccurrence can be adopted by practitioners faced due to lack of concrete guidelines towards sinonasal adenoid cystic carcinoma.

2. Keywords:
Sinonasal adenoid cystic carcinoma;Rare cancer; Endoscopic resection; Case report

3. Introduction and Importance

Adenoid cystic carcinoma is a head and neck cancer that most often involve the salivary glands however sinonasal adenoid cystic carcinoma (SACC) is a possible rare entity[1]. Sinonasal cancers account for just 3% of head and neck cancers with squamous cell carcinoma (SCC) the most common type, followed by adenocarcinoma and then adenoid cystic carcinoma making up less than 10% of sinonasal carcinomas[2]. Histologically they are categorised as tubular, cribriform, solid, and mixed type with solid type associated with worst prognosis with higher rates of invasion and reoccurrence [3]. They tend to present with nonspecific symptoms such as pain, epistaxis, nasal obstruction, and headaches hence tend to be treated for recurrent sinusitis before suspicion for neoplasia arises [4].

They tend to present with widespread local invasion with perineural spread hence the reason for high rates of local reoccurrence despite

extensive surgical resection [5]. Reoccurrence has been seen in tumours with perineural invasion, solid histological type, lymph node metastasis and distant metastasis. Due to propensity for extensive invasion coupled with the rare occurrence of SACC treatment modality of choice is still under discussion. Most institutions however have adopted an aggressive approach towards SACC with extensive surgical resection followed by radiation therapy exhibiting less rates of reoccurrence [6]. Conventional surgery involved an open approach but issues with deformity and reconstruction were raised hence an endoscopic approach has been attempted to achieve tumour free margins to a certain degree [7].

We report a case of a 32-year-old woman who presented with nonspecific nasal symptoms and worked up to have SACC who underwent endoscopic surgical resection followed by cycles of radiotherapy. We describe our experience with diagnosis, oncological work up, surgical resection, radiotherapy and follow up. This paper has been reported in line with the SCARE 2020 criteria [8]. This article has been registered with the Research Registry with identification number researchregistry7949 and can be found through the following hyperlink Browse the Registry - Research Registry.

4. Case Presentation

A 32-year-old female presented to the out-patient ear, nose, and throat (ENT) clinic complaining of left sided nasal pain for 7 months associated with history of recurrent left nasal blockage, headache, and eye pain. No history of epistaxis, change in smell, blurry vision, neither loss of facial sensation or movement nor reported weight loss. Had significant family history of mother having tongue cancer which had metastasised to the neck. She otherwise had no drug allergies, nulliparous and did not smoke or drink alcohol. During the course of illness would be diagnosed as sinusitis on multiple occasions and treated with multiple courses of antibiotics and steroids with minimal relief. On examination she was alert, oriented, and not pale with stable vitals. Had no obvious skin changes or swelling around left nasal region, had nasal blockage with tenderness over maxillary sinus, no cranial nerve palsy noted with intact facial movements and sensation. Rest of systemic examinations were normal.

On initial evaluation Computed tomography (CT) of the Paranasal Sinuses (PNS) revealed homogeneous opacification of the left maxillary sinus, ethmoidal air cells and frontal sinus with expansion of the left maxillary sinus accompanied with thinning of its medial wall and bulging of medial wall into the left nasal cavity causing obstruction (Fig 1). Suspicion for a possible mucocele, polyp and neoplasm were raised hence an endoscopic sinus evaluation carried out by a senior ENT surgeon revealed an eroded medial wall of the maxillary sinus with a fungating and friable mass arising from the maxillary and ethmoid sinus which bled easily along with pus collection within the maxillary sinus.

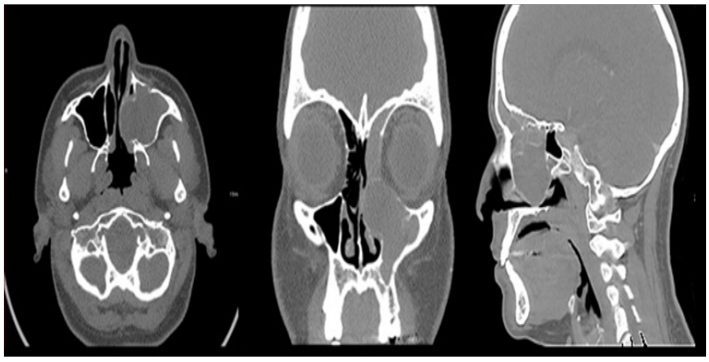

Fig 1: Pre contrast CT PNS (Axial, Coronal and Sagittal Views in Bone window) showing expansile homogeneous non-contrast enhancing opacification of left maxillary sinus with thinning/erosion of the medial wall and opacification of ethmoidal air cells and frontal sinus.

At the same sitting underwent endoscopic medial maxillectomy plus anterior and posterior ethmoidectomy with tissue sent for histopathology analysis which revealed a maxillary sinus adenoid cystic carcinoma without perineural invasion (Fig 2). The sections showed a malignant neoplasm arising from submucosa glands of maxillary sinus and the pattern was mostly cribriform with some areas showing tubular formation. There were cystic spaces containing mucoid material and tumour cells were small uniform cuboidal with scant cytoplasm and rare mitosis. The tumour extended to the surrounding bone but perineural invasion was not identified.

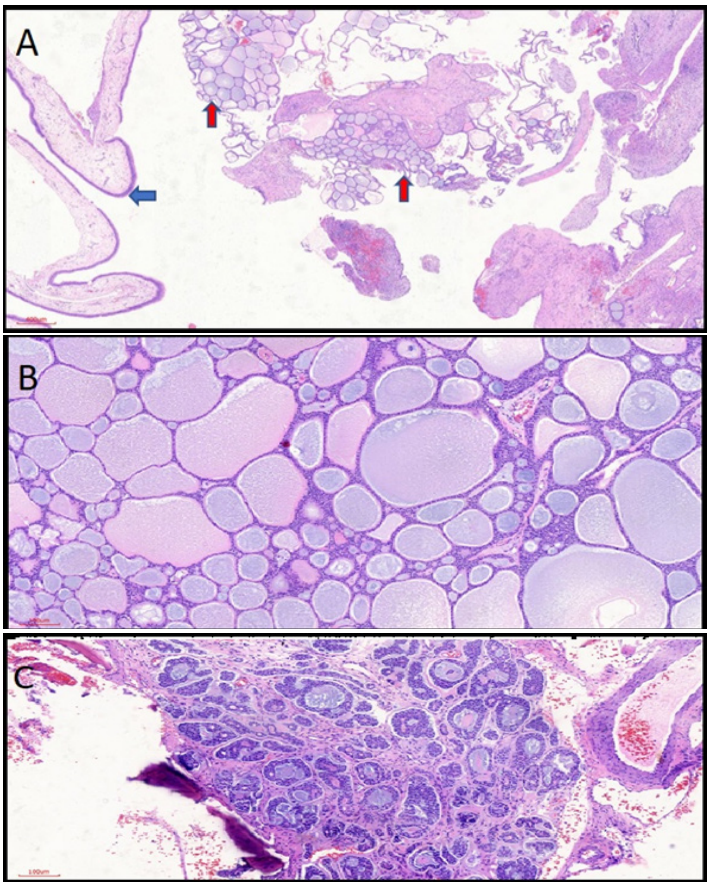

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Fig 2: A – Low power view; Hematoxylin and Eosin sections showing a tumour in cribriform pattern on the middle of the image (red arrows) and left side showing maxillary sinus epithelial lining (blue arrow). B – High power view; Hematoxylin and Eosin sections showing mostly cribriform pattern with cystic spaces filled with mucoid materials and surrounded by uniform cuboidal tumor cells. C – A low power view; Hematoxylin and Eosin showing small myoepithelial cells with bland cytology surrounding pseudoglandular structures containing mucin as well as tubular structures which were extending to the adjacent bone (on the left side)

Case was discussed at the tumour board and a multidisciplinary approach carried out with metastatic work up revealing no extension into the base of skull, brain, nerves, or neck without any distant metastasis. However Magnetic resonance imaging (MRI) of the PNS revealed an area with abnormal fluid restriction within the left anterior ethmoid sinus suggestive of residual tumour as well as mucosal thickening in the left maxillary, ethmoidal and frontal sinuses (Fig 3). Therefore, considering her age with functional and cosmetic outcome, she underwent a functional endoscopic sinus surgery to resect the residual tumour until tumour free margins were achieved and confirmed on frozen section analysis. Histopathology showed deposits of SACC in tissue lateral and posterior to nasolacrimal duct and tissue along the orbital floor with negative margins and no invasion into perineural and lymphovascular structures. Cerebrospinal fluid analysis was normal without presence of malignant cells on cytology analysis.

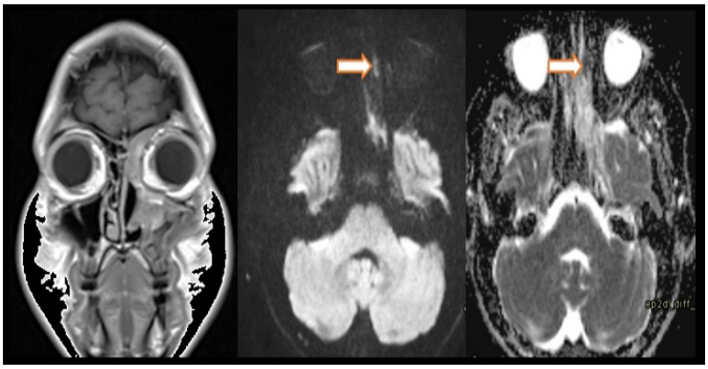

Fig 3: MRI TIWI Coronal, Axial DWI, and ADC Images – MRI Post surgery revealed diffuse mucosal thickening in the left maxillary sinus, left ethmoidal air cells and left frontal sinus seen on the T1WI coronal view and abnormal fluid restriction in the left ethmoid air cell on DWI with corresponding low signal on ADC.

Post-operative the patient received external beam radiation therapy as part of curative intent at 2Gy per cycle for 32 cycles for a total radiation dose of 64Gy. Upon completion of radiotherapy follow up MRI at 3 months revealed no areas of fluid restriction to suggest no residual tumour with diffuse mucosal thickening to suggest chronic sinusitis (Fig 4). Due to lack of availability at 6 months post radiation patient travelled overseas for a Positron Emission Tomography (PET) scan which revealed findings suggestive of chronic inflammation without residual tumour. Is now 2 years post radiation and has currently resumed to daily activities with complains of on and off headaches and episodes of nasal congestion. Thus, is on symptomatic treatment with flupiritine and nasal douching with saline drops as per need basis with a scheduled follow up plan to assess for reoccurrence in place as per National Comprehensive Cancer Network guidelines.

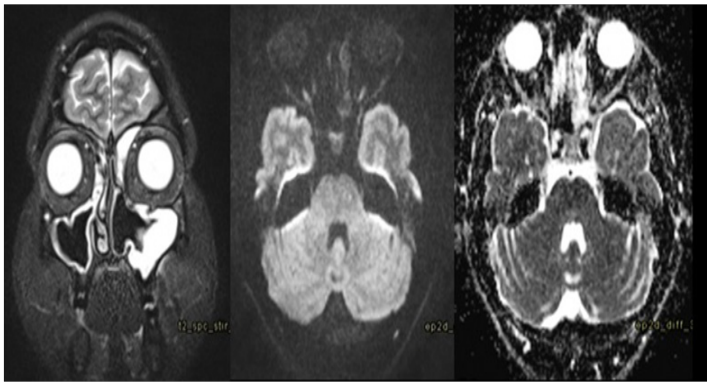

Fig 4: MRI T2 Stir Coronal, Axial Diffusion and ADC Images – MRI 3 months post FESS and radiotherapy revealedMRI revaled diffuse mucosal thickening to suggest chronic sinusitis on T2 Stir coronal image and no areas of fluid restriction to suggest absence residual tumour.

5. Discussion

Clinical manifestations initially are very nonspecific with epistaxis and nasal obstruction with prior history of treatment for recurrent sinusitis without resolution of symptoms. Similarly, in our case our patient bounced from multiple outpatient clinics and was given treatment for sinusitis however symptoms did not resolve. Upon spread of the carcinoma symptoms range from pain due to bone involvement to symptoms of nerve palsy due to perineural invasion [9]. Therefore, we performed a thorough neurological examination as warranted in patients with SACC with particular emphasis towards cranial nerve examination however did not reveal any features of neural involvement which was confirmed on histological analysis.

Most sinonasal carcinomas are predominantly SCC followed by adenocarcinomas with SACC constituting a rare form of sinonasal carcinoma[10]. They are slow growing however have tendencies towards perineural invasion along with local invasion involving bone and in extreme cases intracranial extension reported in isolated case reports[11, 12]. Due to the local infiltration extensive surgical resection is the mainstay of treatment followed by radiotherapy to reduce risk of reoccurrence [13]. As with our case the patient underwent surgical resection to achieve tumour free margins followed by radiotherapy sessions with follow up imaging revealing complete excision without sites of reoccurrence on follow up imaging. Debate has been raised on the efficacy of an endoscopic approach for oncological resection of SACC due to lack of larger studies due to the rarity of the cancer. Studies however comparing endoscopic resection to

open resection for other forms of PNS tumours have been carried out. Endoscopic approach for sinonasal melanoma had comparable outcomes to open surgery for proportion of negative margins, median survival, and disease-free survival rates [14]. However endoscopic approach also offered the attractive option of minimal invasive with lack of scar and deformity needing reconstruction. Endoscopic approach in well selected patients has shown to be effective in achieving acceptable oncological outcomes for sinonasal carcinomas with a 5 and 10-year disease-specific survival rates of 87% and 80%, respectively. It provides satisfactory survival rates with better post-operative quality of life, better aesthetic outcome and reduces the need for craniofacial dissection with or without reconstruction[15, 16].

CT scan has shown great sensitivity, specificity, and accuracy in number of different sinonasal diseases such as; inflammatory diseases, infections and neoplasms causing clinical diagnostic dilemma as the signs and symptoms are non-specific. CT scan can demonstrate the extent of the sinonasal diseases as well as its impact on the adjacent bones i.e., expansion, thinning or erosion. However, in some clinical settings, even sophisticated CT scan is unable to sufficiently characterise the sinonasal lesions and so MRI may be required for further characterization. In this regard, patients' CT scan findings must also be correlated with cytohistopathology. MRI of the brain and PNS can reveal the infiltrative nature of the SACC and the natural tendency to spread along the perineural spaces into the skull base. MRI offers a better differentiation of tumour from surrounding tissues and post-surgical evaluation is achieved multiplane and multi-sequential pre and post contrast MRI.

Conclusions on the gold standard therapeutic approach are still under debate however working up patients with suspicion for sinonasal mass regardless of histological type involves a CT of the PNS [3]. Due to suspicion for a possible sinonasal carcinoma in our case after CT imaging an endoscopic evaluation revealed a SACC hence an MRI to identify for local invasion into cranial or neurological structures. Post-operative imaging at 3 months follows up after resection and radiotherapy to assess for reoccurrence was successfully concluded upon via an MRI and a 6-month imaging follow up via PET CT scan. To our knowledge this is the only reported case report of SACC from East Africa stressing on the rarity of SACC as well as underdiagnoses due to the tendency to be treated as a sinusitis because of nonspecific symptoms. Through our experience via an endoscopic approach, we endorse the recommendation for SACC to undergo surgical resection with endoscopy as a possible route to achieve tumour free margins followed by radiotherapy to prevent reoccurrence despite its tendency to reoccur with a close follow up program in place.

6. Conclusion

Sinonasal adenoid cystic carcinoma is a rare carcinoma of the sinonasal region presenting with nonspecific nasal symptoms. We hope through our successful experience of achieving adequate resection via an endoscopic approach followed by radiotherapy to prevent reoccurrence can be adopted by practitioners faced with a SACC due to lack of concrete guidelines towards it. We are aware of the fact that the assessment of survival in this case is inappropriate in over short period however, the objective of the case was to emphasize the usefulness of endoscopic approach in minimal invasive removal of SNACC as well as to raise awareness in patients with recurrent sinusitis not responding to therapy for possible catastrophic disease.

7. Abbreviations:

ADC – Apparent Diffusion Coefficient, CT – Computed Tomography, DWI – Diffusion Weighted Image, PNS – Paranasal Sinus, PET – Positron Emission Tomography, MRI – Magnetic Resonance Imaging, SACC – SinonasalAdenoid Cystic Carcinoma, SCC – Squamous Cell Carnicoma.

8. Patient's perspective:

I was surprised when I was told I had a cancer as I have had nasal symptoms for a long time. Undergoing surgery and radiation was overwhelming however I am incredibly pleased with my most recent imaging revealing there was no reoccurrence of the cancer.

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