Case study

Angiomatoid Fibrous Histiocytoma of the Neck Mimicking a Large Nodal Metastatic Carcinoma: A rare tumour at an unusual site

ABSTRACT

Aims: Angiomatoid fibrous histiocytoma, a tumour of uncertain differentiation and intermediate behaviour, is rarely seen in the head and neck region. We report its dramatic presentation in the neck as a progressive, large mass mimicking malignancy clinically as well as cytologically, to share our challenges in diagnosing and treating this condition.
Presentation of case: A healthy 40-year-old gentleman presented to our centre with a 2-year history of a slowly progressive large right neck mass, without any constitutional symptoms. Examination showed a large right neck mass, and subsequent fine needle aspiration revealed metastatic carcinoma. Loss in follow up, he returned 2 years later without any progression. This unusual behaviour raised suspicion, and subsequent incisional biopsy revealed a spindle cell neoplasm suggestive of a sarcoma. Excision of the lesion showed a well-encapsulated tumour, and subsequent histopathological examination revealed an angiomatoid fibrous histiocytoma of the neck.
Discussion: Angiomatoid fibrous histiocytoma presents a diagnostic dilemma to clinicians as: clinically, it may display similar appearance to a lipoma, haematoma or haemangioma; and pathological examination of the lesion may mimic metastatic carcinoma or melanoma, even in the presence of distinctive morphology. The mainstay of treatment is surgical excision, and in the rare case of recurrence or metastasis; chemoradiotherapy.
Conclusion: A slowly progressive neck mass reported as metastatic carcinoma should be investigated with caution

Keywords: Angiomatoid fibrous histiocytoma, neck, neck masses, cervical lymph node metastases

1. INTRODUCTION

Angiomatoid fibrous histiocytoma (AFH) is a rare subcutaneous soft tissue tumour of uncertain differentiation, accounting for only 0.3% of all soft tissue tumours. The head and neck region is an uncommon site for the tumour, amounting to only 7% of AFH, which is usually found in trunks or the extremities. (1, 2) Initially described by Enzinger as malignant, this tumour is now known to exhibit an intermediate behaviour with a low rate of recurrence and metastasis (up to 11% and less than 5%, respectively), and rarely resulting in death. (1, 3)

2. PRESENTTION OF CASE

A healthy, 41-year-old male construction technician presented to our outpatient clinic with 2-year-history of a painless but slowly progressive right neck mass. There was no history of preceding injury, trauma or infection. Examination revealed a large solitary firm mass at the right lateral neck measuring 10 x 12 centimetres, which appeared to be deep-seated. Fine needle aspiration cytological examination (FNAC) of the mass showed atypical epithelial cells with enlarged, pleomorphic and irregular nuclei in a lymphoid background, which were consistent with metastatic carcinoma in a lymph node. However, subsequent panendoscopic examination of the upper aerodigestive tract and upper and lower gastrointestinal endoscopy did not reveal any detectable primary. Computerised tomography (CT) showed a large solitary well-defined, poorly enhancing, oval-shaped homogenous soft tissue mass. However, the rest of the thorax, abdomen and pelvis were normal.

A neck dissection was planned, however, the patient defaulted and returned a year later with only a slight increase in the size of the neck mass without any other new symptoms. Given the initial diagnosis, the absence of aggressive behaviour of the tumour raised suspicion amongst the clinicians treating him. Thus an incisional biopsy was performed, which showed uniform ovoid cells in syncytial sheets, suggestive of a spindle cell neoplasm. An excision of the neck mass was then planned, however after coming back with an episode of bleeding from the biopsy site, the patient subsequently did not attend further follow-up. Amazingly, he returned 2 years later with an increase of the lesion size without any other progression, which was evident on repeat CT. We subsequently proceeded with excision of the tumour – intraoperatively, it was a well-encapsulated tumour without any gross invasion of surrounding structures. Excision was uneventful, and the patient recovered well postoperatively. Macroscopic pathological examination showed a fully capsulated and bosselated greyish mass weighing 400 grams and measuring 13 cm x 9 cm x 8 cm, with blood-filled spaces on cut sections. Microscopically, the tumour was composed of syncytial sheets of histiocytoid cells, interspersed with numerous, dilated, psudoangiomatoid spaces, with prominent lymphoplasmatic infiltrate, all within a thick fibrocollagenous capsule. Immunohistochemistry tests were positive for C68 and CD99, and negative for SMA and Desmin with a low proliferative index for Ki67. All the margins were clear. The histopathological examination of the specimen was consistent with angiomatoid fibrous histiocytoma.

3. DISCUSSION

AFH usually presents as a slowly-growing painless tumour of the deep dermis and subcutis, and is frequently associated with an antecedent trauma to the area. (2) It is rarely seen in the neck, and to the author's knowledge, there is only one report available in the literature. (4) In addition to the fact that most of AFH does not occur in areas where lymph nodes are found, its non-specific clinical feature may lead to initial misdiagnosis as haematoma, haemangioma or even lipoma. (1) Our case demonstrated a somewhat atypical presentation of the lesion as a firm, fixed and deep-seated mass located at the neck, much more akin to a suspicious lymphadenopathy.

With regards to imaging modalities, CT of AFH may show lytic lesions with thin, enhancing septations; however these are often, as with our case, non-specific. (5) MRI may be more specific, as reported in a case series, in which subjects with AFH were found to share characteristic findings of a cystic solid mass with a double rim sign, as well as evidence of infiltrating margins. (6) Interestingly, a report described the ultrasonographic findings of AFH

that correlated clearly with its histological characteristics, suggesting that sonographic imaging may in fact be more useful than previously thought in diagnosing AFH; this was further supported in another, more recent case series. (7, 8)

Histologically, typical AFH is characterised by 4 key features, with at least 2 present in most cases: (i) syncytial sheets of spindle or oval histiocytoid cells; (ii) pseudoangiomatoid spaces; (iii) a thick fibrous capsule; and (iv) a prominent lymphoplasmacytic infiltrate. (2, 9) However, a broad spectrum of histopathological and cytological findings exists, which may result in diagnostic pitfalls. (9) In particular, the presence of cellular atypia and more commonly nuclear pleomorphism in some cases may mimic carcinoma, which indeed was the case in the cytological sample of this patient's lesion; and without a repeat sample, he might have been subjected to the possible complications of an unnecessary neck dissection. Additionally, AFH also lacks an immunoprofile, and while molecular genetic studies have shown characteristic translocations (such as *EWSR1-CREB1*), these were not unique to it. (2, 9) Overall, the main challenge for us was the dilemma faced in accurately diagnosing this condition preoperatively, which was attributed to the rarity of its incidence as well as its histological spectrum.

Most AFH behave indolently with relatively low rates of recurrence and metastasis, and the appropriate treatment for AFH is wide local excision with follow up for recurrence or metastasis. (4) The likelihood of the tumour to recur or metastasise has been found to correlate with invasion into deep fascia or muscle, but not to various histologic parameters such as mitotic activity or pleomorphism. Additionally, the location of the tumour in the head and neck region is associated with a higher possibility of local recurrence, likely due to the difficulty in performing a wide local excision in this region. (3) Local recurrence may be amenable to excision however in cases where excision is not feasible, or if regional nodes are involved, chemotherapy have been shown to be effective. (10, 11) While the histopathological examination of the tumour margins in our patient was reassuring, its location in the neck warranted follow-up, and we report our patient remains free of recurrence 2 years after complete excision.

4. CONCLUSION

Angiomatoid fibrous histiocytoma in the neck is rare and may mimic other pathologies. A slowly-progressing neck mass that is reported as metastatic carcinoma should be investigated with caution to avoid unnecessary radical surgery. Ultrasonography may prove to be an indispensable tool in diagnosing the tumour.

COMPETING INTERESTS

None declared

CONSENT

All authors declare that written informed consent was obtained from the patient for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editorial office/Chief Editor/Editorial Board members of this journal.

ETHICAL APPROVAL

As per international standard written ethical approval has been collected and preserved by the author(s).

REFERENCES

1. Enzinger FM. Angiomatoid malignant fibrous histiocytoma: a distinct fibrohistiocytic tumor of children and young adults simulating a vascular neoplasm. Cancer. 1979;44(6):2147-57.
2. Fanburg-Smith JC, Dal Chin P. Angiomatoid Fibrous Histiocytoma. In: Fletcher CDM. UK, Mertens F. (Eds), editor. World Health Organization Classification of Tumours Pathology and Genetics of Tumours of Soft Tissue and Bone. Lyon, France: IARC Press; 2002. p. 194-5.
3. Costa MJ, Weiss SW. Angiomatoid malignant fibrous histiocytoma. A follow-up study of 108 cases with evaluation of possible histologic predictors of outcome. Am J Surg Pathol. 1990;14(12):1126-32.
4. Lai EC, Chung KM, Chiu HF, Lau WY. Angiomatoid fibrous histiocytoma in the neck. ANZ journal of surgery. 2006;76(6):538.
5. Petrey WB, LeGallo RD, Fox MG, Gaskin CM. Imaging characteristics of angiomatoid fibrous histiocytoma of bone. Skeletal Radiol. 2011;40(2):233-7.
6. Martinez SJ, Moreno CC, Vinson EN, Dodd LG, Brigman BE. Angiomatoid fibrous histiocytoma: novel MR imaging findings. Skeletal Radiol. 2016;45(5):661-70.
7. Hata H, Natsuga K, Aoyagi S, Homma E, Shimizu H. Ultrasound B-mode and elastographic findings of angiomatoid fibrous histiocytoma. Clin Exp Dermatol. 2014;39(4):538-9.
8. Yikilmaz A, Ngan BY, Navarro OM. Imaging of childhood angiomatoid fibrous histiocytoma with pathological correlation. Pediatr Radiol. 2015;45(12):1796-802.
9. Thway K, Fisher C. Angiomatoid fibrous histiocytoma: the current status of pathology and genetics. Arch Pathol Lab Med. 2015;139(5):674-82.

10. Ogden S, Harave S, McPartland J, Brennan B, Jeys L, Losty P, et al. Angiomatoid fibrous histiocytoma: A case of local recurrence and metastases to loco-regional lymph nodes that responded to chemotherapy. Pediatr Blood Cancer. 2016.
11. Costa MA, Silva I, Carvalhido L, Azevedo I, Alves L, Leal C, et al. Angiomatoid fibrous histiocytoma of the arm treated by radiotherapy for local recurrence--case report. Med Pediatr Oncol. 1997;28(5):373-6.

APPENDIX

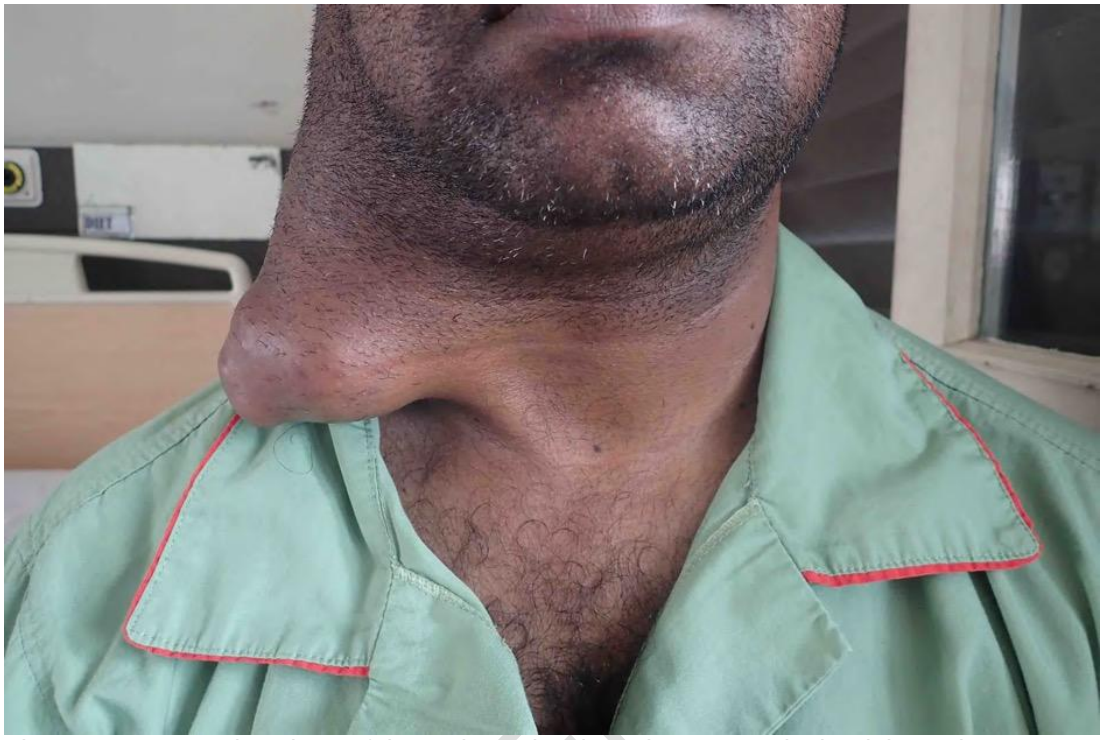

Figure 1: Preoperative photo of the patient, showing a large mass in the right neck

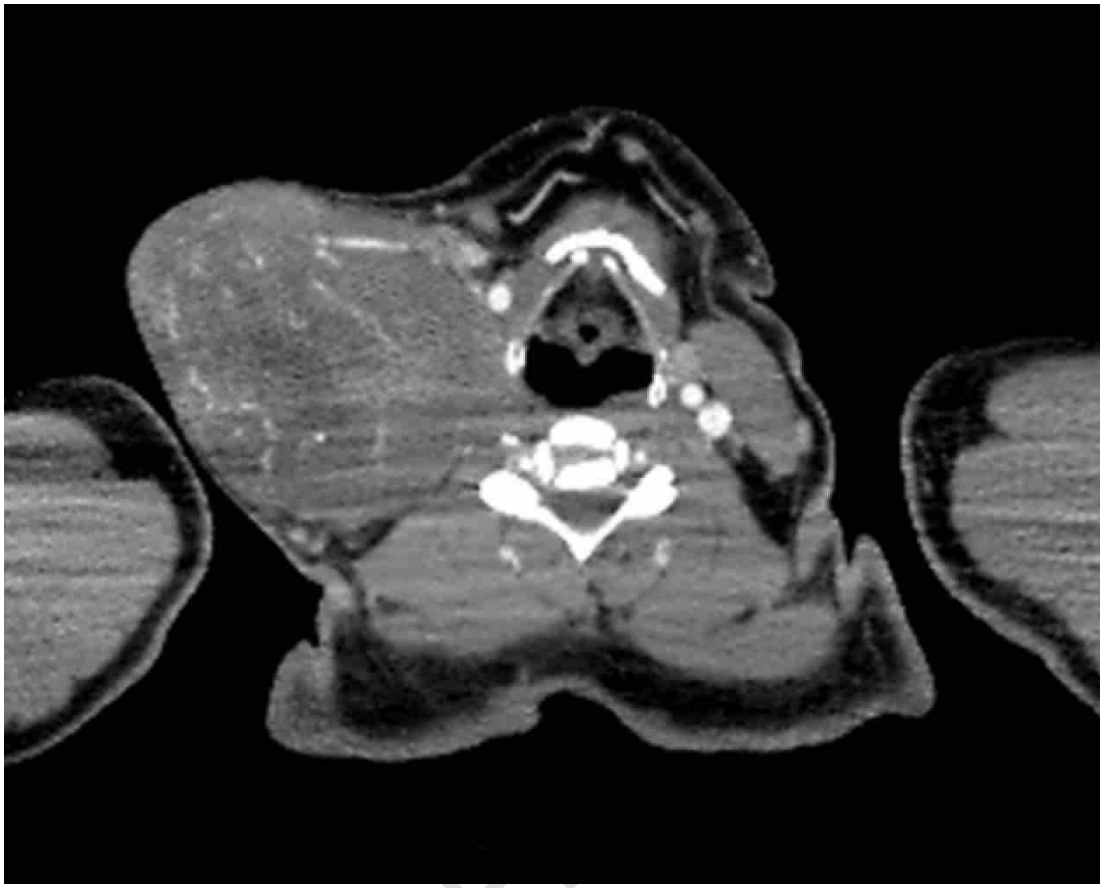

Figure 2: A contrast enhanced computer tomography of the neck in axial view, showing the neck mass with increased vascularity

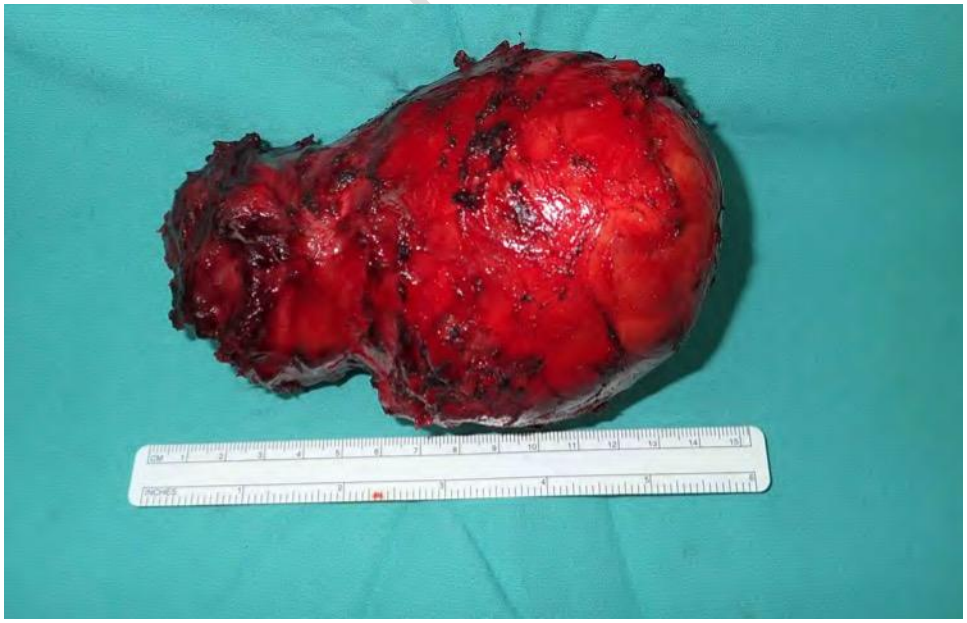

Figure 3: Excised tumour appeared well encapsulated

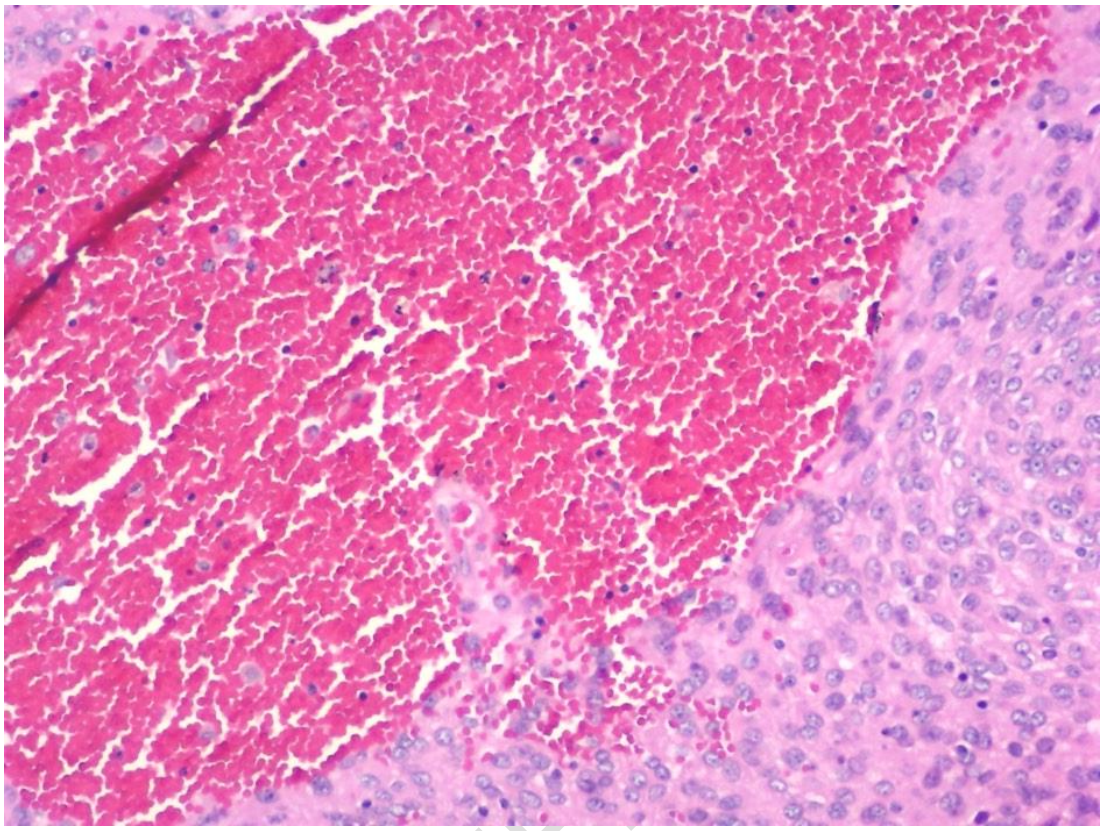

Figure 4: Dilated spaced containing blood and devoid of lining epithelium (H&E, 10x)

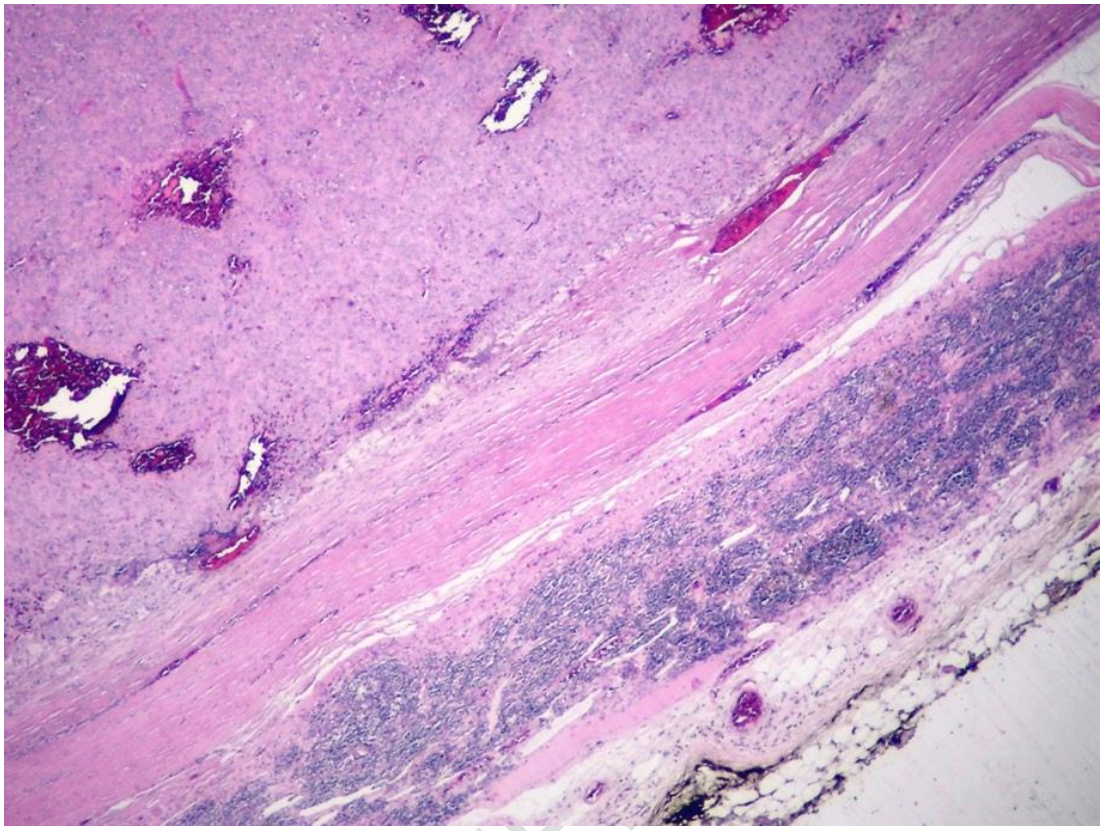

Figure 5: Prominent lymphoplasmacytic infiltrate with occasional germinal centre formation are seen within the mass and at the subcapsular area (H&E, 2x)

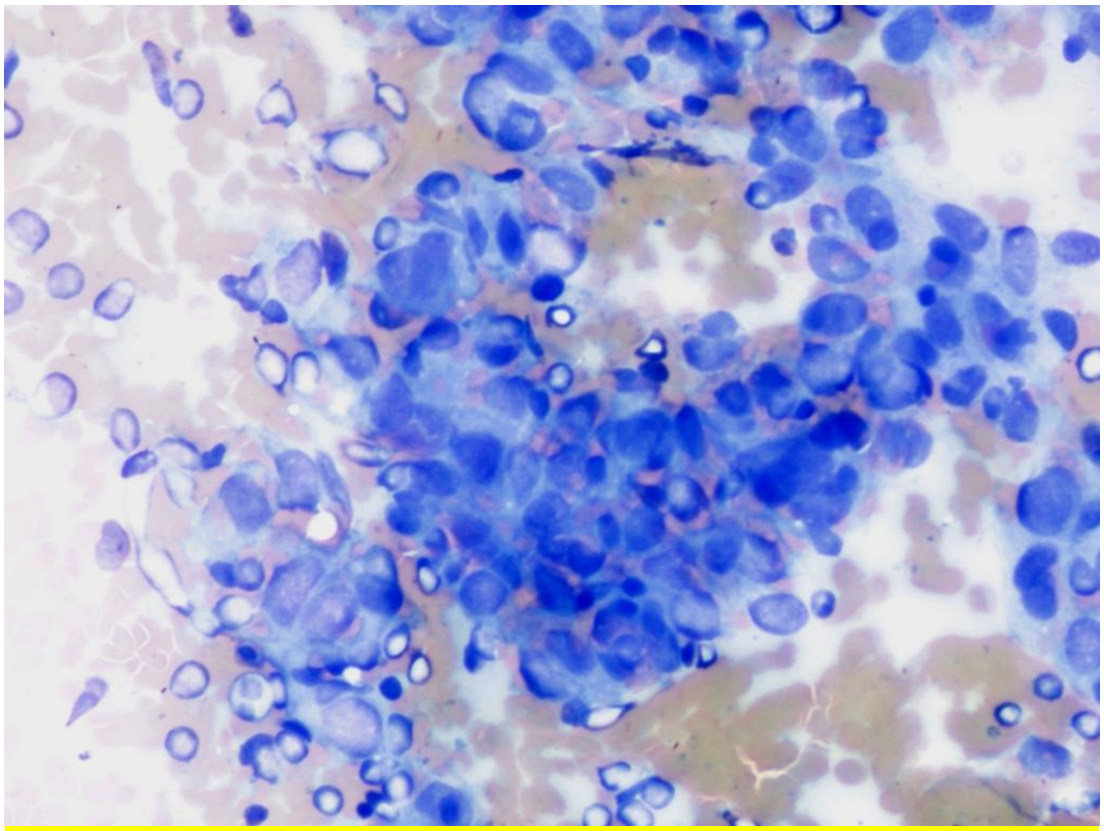

Figure 6: A cluster of atypical cells in the fine needle aspirate showing enlarged, pleomorphic and irregular nuclei with ample cytoplasm (MGG, 20x).