

The Gulf Journal of Oncology



Indexed By PubMed and Medline Database

Issue 44, January 2024

ISSN No. 2078-2101



70th session of the WHO Regional Committee for the Eastern Mediterranean, Cairo, Egypt 9-12 October 2023



الأسبوع الخليجي التاسع للتوعية بالسرطان
9th Gulf Week for Cancer Awareness

1-7 February 2024



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1937-2023

The Official Journal of the Gulf Federation For Cancer Control

Table of Contents

Original Articles

Assessment of Uncertainty in Volume Estimation of Non–Static Target: A Phantom Study using Racemosa Wood	07
TR Verma, NK Painuly, SP Mishar, MLB Bhatt	
Immunohistopathological Study of Papillary Squamotransitional Carcinoma of Uterine Cervix	16
Bhagyashree Dhande, Siddhi Gaurish Sinai Khandeparkar, Bageshri P. Gogate, Avinash R. Joshi, Shital Subhash Gosavi, Pooja Vinod Mishraa	
Clinical and Pathological Characteristics of Breast Cancer Among Emirati National Patients.....	25
Rafal R. Iskanderian, Ahmed Matakah, Aya Abdoh, Asma Al Hashmi, Bassel Jallad, Fady Geara, Stephen R. Grobmyer	
Evaluation of Dentist’s Knowledge and Practice About Dental Management of Oral Cancer Patients.....	30
Faezeh khozeimeh, Hossein Hadi, Hanieh Haghpanah, Bahareh Tahani	
A Prospective Study of the Incidence of Chronic Xerostomia and the Quality of Life in Patients Undergoing Radiotherapy for Head and Neck Malignancies with IMRT or VMAT Techniques	39
Nishant Gaurav Pathak, Mahadev P, Yash Alok	
URS–SM procedure using Pneumatic lithotripter for the management of ureteral stones–our experience	48
Xh. Çuni, S. Mehmeti, A. Neziri, L. Çuni, D. Mucaj, D. Çuni	
Long–term Outcomes of Cancer Patients Admitted to the ICU with Septic Shock.....	51
Wedad B. Awad, Lama Nazer	

Review Article

Physiotherapy in Head, Neck, Lung and Breast Cancer Survivors: A Systematic Review.....	54
Fatima Abdul Rashid, Wajiha Anwar, Samiya Malik, Meruna Bose, Praveen Kumar, Animesh Hazari	
Trends in Stereotactic Radiosurgery for Meningioma: the Top 100 Most Cited Articles	66
Tariq Al Habsi, Hashim Alibrahim, Utba Al Manthari, Adham Al–Rahbi, Tariq Al–Saadi	
Biomarkers in Prostate Cancer: A Review.....	81
Nizar Ahmadih, Toufic Zeidan, Celine Chaaya, David Cain, Marc Aoude, Anita Abouchahla, Hampig Raphael Kourie, Elie Nemer	
Synergizing Expertise and Technology: The Artificial intelligence Revolution in Radiotherapy for Personalized and Precise Cancer Treatment.....	94
Fadila Kouhen, Hanae EL Gouach, Kamal Saidi, Zineb Dahbi, Nadia Errafiy, Hafsa Elmarrachi, Nabil Ismaili	

Case Report

Clear Cell Meningioma with Tyrosine Crystals: a Case Report with Review of Literature	103
Rashim Sharma, Poonam Abhay Elhence, Deepak Vedant, Jigish Ruparelia, Suryanarayanan Bhaskar, Sarbesh Tiwari	

Conference Highlights/Scientific Contributions

• News Notes.....	107
• Advertisements	111
• Scientific Activities	112



Case Report

Clear Cell Meningioma with Tyrosine Crystals: a Case Report with Review of Literature

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Abstract

Introduction or Background: Clear cell meningioma is a rare WHO grade 2 tumour and runs an aggressive course. Tyrosine crystals are very uncommon in meningioma.

Case Presentation: We present a case of a 43-year female with right middle and posterior cranial fossa space occupying lesion (SOL) diagnosed as clear cell

meningioma with presence of numerous tyrosine crystals.

Discussion and Conclusion: Whether these crystals are incidentally noted or if there is an actual relationship of these crystals with tumour environment must be found as they are seen in many other conditions too.

Keywords: Tumour, Meningioma, Central nervous system, Tyrosine

Introduction

Meningioma accounts for one third of all primary central nervous system neoplasms.⁽¹⁾ Clear cell meningioma is a rare WHO Grade 2 tumour which forms 0.2–0.8% of all the meningiomas and has an aggressive behaviour.^(2,3) Tyrosine crystals are rarely seen in meningioma and there are scarce cases reported in the literature. We present a case of clear cell meningioma in a 43-year female with the presence of numerous tyrosine crystals.

Case Presentation

A 43-year-old female presented with complaints of severe headache and decreased hearing for 6 months along with difficulty in walking for 1 month. There was no history of vision loss, fever or any chronic illness. Clinical examination was unremarkable. Non contrast CT head showed an ill-defined hyperattenuating extra-axial mass epicentered at the right prepontine and right cerebellopontine angle cistern extending to right middle cranial fossa (Figure 1A). T2W MR brain showed the lesion to be heterogeneous in signal intensity with internal cysts and epicentered at right petroclinoid ligament (Figure 1B). Susceptibility weighted imaging didn't show any obvious haemorrhage or calcification within (Figure 1C). The ADC map of diffusion images showed few foci of restriction within (Figure 1D). The post contrast scan revealed heterogeneous moderate enhancement with areas of

internal necrosis (Figure 1E). The cerebral blood volume perfusion image shows the lesion to be hypoperfused (Figure 1E). A right temporal craniotomy was performed and intra-operatively, tumour was whitish, soft to firm with cystic spaces. Tumour was only mildly vascular, not easily suckable.

On light microscopy, lobules and focal whorls of tumour cells were noted. The tumour cells were polyhedral with round monomorphic to few enlarged nuclei, occasional punctate to conspicuous nucleoli and moderate amounts of well-defined clear PAS positive cytoplasm. Abundant variably shaped interstitial collagen was noted with numerous daisy head-like crystals highlighting on Hematoxylin and Eosin stain (H&E) and Masson's Trichrome stain (MT) (Figure 2 A, B). Mitosis was occasionally seen. No necrosis, psammoma bodies or intranuclear inclusions were seen.

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On Immunohistochemistry, the tumour cells were positive for Progesterone receptor (PgR), Vimentin (Figure 3 A, B) and Epithelial membrane antigen (EMA). The tumour cells were negative for S100 protein. Ki-67 labelling index was 3–4%. Diagnosis of Clear cell meningioma, WHO grade 2 was given. The patient is doing well and is on follow up.

Discussion and Conclusion

Manivel and Sung in late 1990's coined the term Clear cell meningioma.⁽⁴⁾ It has an affinity for cerebellopontine angle, similar to our case and affects younger age group.⁽⁵⁾ On histomorphology, it has patternless arrangement or can be in sheets. The cells are round to polygonal and

have a regular cytoplasmic membrane with abundant clear, glycogenated cytoplasm, round small nucleus, fine chromatin and inconspicuous nucleoli. There is prominence of collagen, both perivascularly and interstitially.⁽⁵⁾ Glycogen stains positive for periodic acid Schiff (PAS) stain. Tyrosine crystals in meningioma are rarely noted. After a thorough search on Pubmed, Cochrane and EMBASE libraries, so far only three cases were found to have reported these findings. Of these three cases, only one case by Schollenberg et al showed tyrosine crystals in clear cell meningioma. The other two cases comprised of atypical meningioma with tyrosine crystals and fibrous meningioma with tyrosine crystals. A review of literature has been tabulated in chronological order in Table 1.^(6–8)

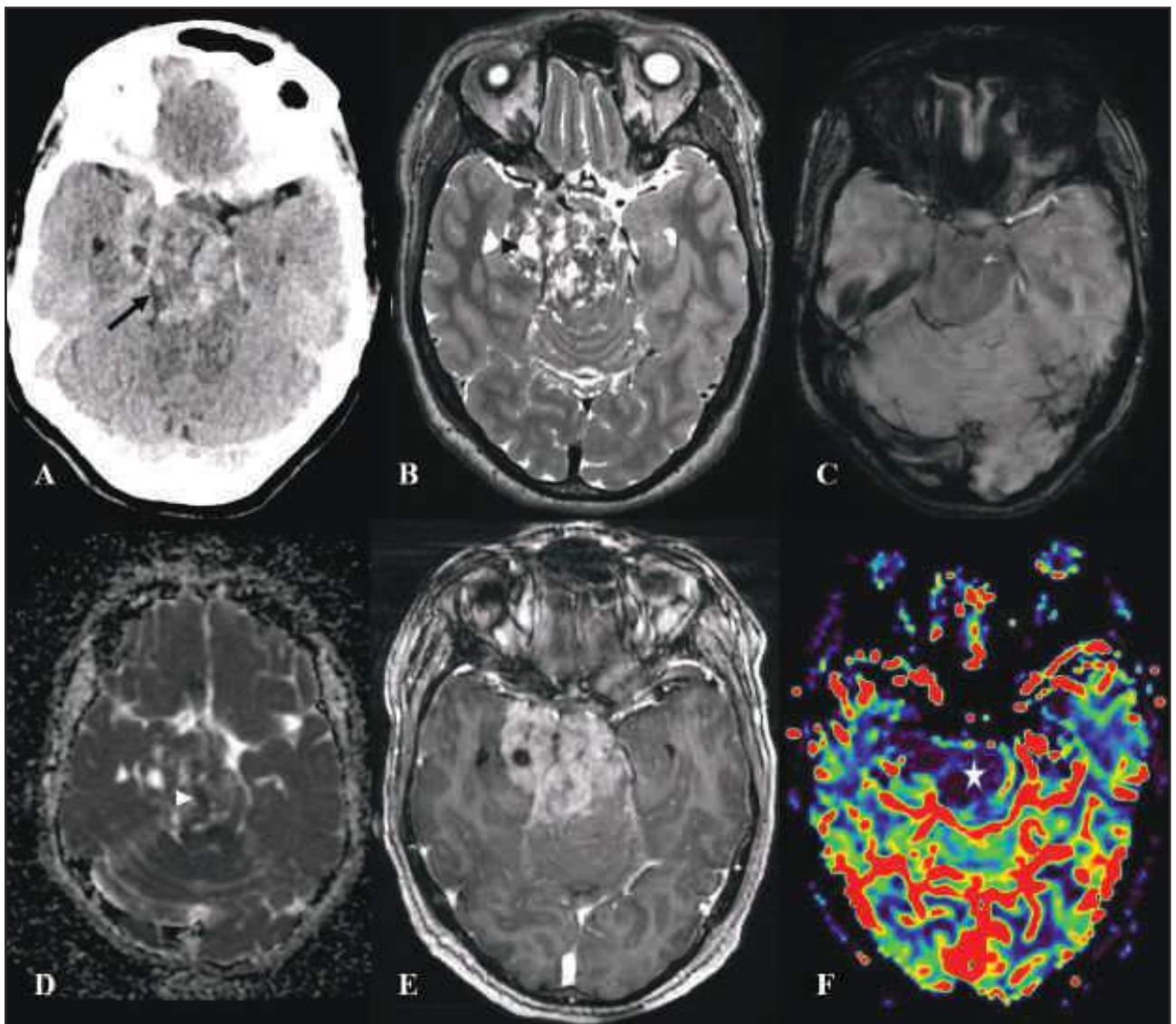


Figure 1. The axial non contrast CT head image (A) shows an ill–defined hyperattenuating extra–axial mass epicentered at the right prepontine and right cerebellopontine angle cistern extending to right middle cranial fossa (black arrow). The axial T2W MR brain (B) shows the lesion to be heterogeneous in signal intensity with internal cysts (black arrowhead) and epicentered at right petroclivoid ligament. The susceptibility weighted imaging (C) didn't show any obvious haemorrhage or calcification within. The ADC map of diffusion images (D) shows few foci of restricted within (white arrowhead). The post contrast scan reveals heterogeneous moderate enhancement (E) with areas of internal necrosis. The cerebral blood volume perfusion image (F) shows the lesion to be hypoperfused (asterix).

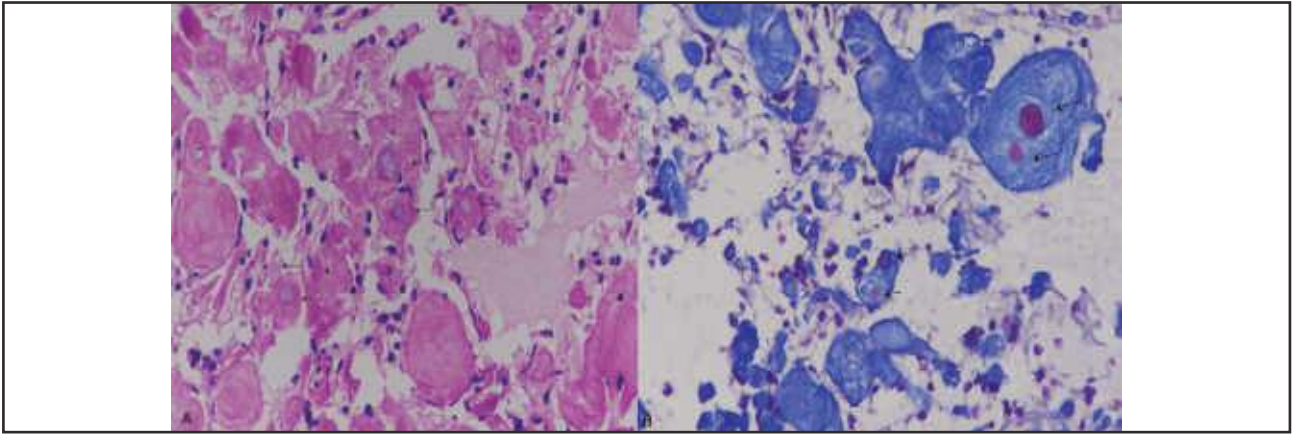


Figure 2. Microscopy images (A) shows numerous daisy head like tyrosine crystals (black arrows) in interspersed meningothelial cells exhibiting clear abundant cytoplasm, round nuclei with fine chromatin and inconspicuous nucleoli (H&E, 40x) (B) shows numerous daisy head like tyrosine crystals (black arrows) (Masson Trichrome stain, 40x)

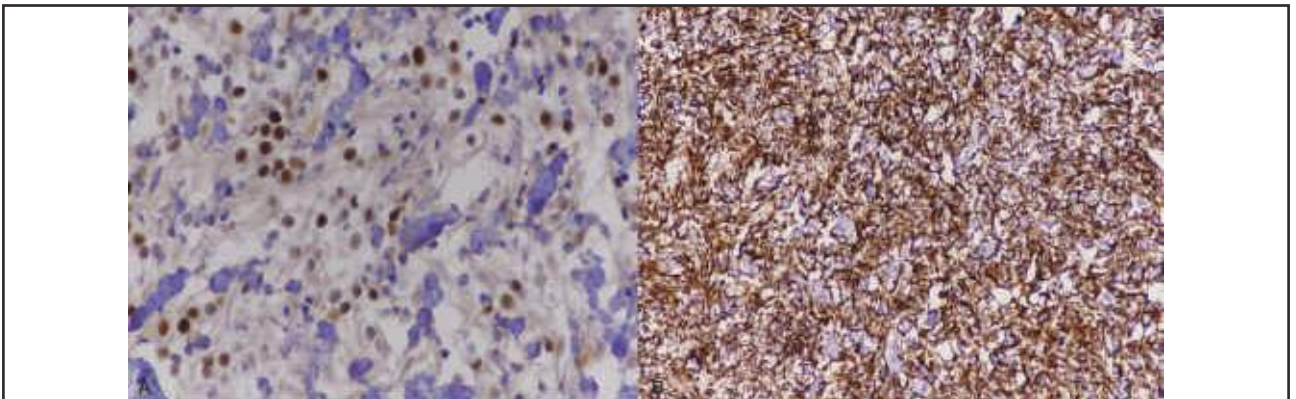


Figure 3. Immunohistochemistry images (A) shows strong and diffuse nuclear positivity for progesterone receptor (PgR) (B) shows strong and diffuse cytoplasmic positivity for Vimentin.

Tyrosine crystals have been noted in salivary gland tumours like, pleomorphic adenoma, polymorphous adenocarcinoma, myoepithelioma and adenoid cystic carcinoma.⁽⁹⁾ These crystals have also been noted in head and neck neoplasms, especially in larynx adjacent to squamous cell carcinoma.⁽¹⁰⁾ These crystals have daisy head or petaloid shape on light microscopy and do not show any birefringence on polarizer.⁽⁶⁾ Presence of tyrosine crystals must be documented. Whether the formation of these crystals is serendipitous or has some contribution to the disease process is not known. However, as these are found in many conditions, a possible explanation must be sought.

Acknowledgement:

None

Funding and Conflict of Interest

No funding received. There is no conflict of interest.

Ethics approval:

Institutional Ethics Committee approval is not warranted for this.

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Authors	Name of study	Year of publication	Clinical findings	Imaging findings	Histopathological diagnosis	Electron microscopy	Other salient findings
Couce et al ^[6]	Fibrous Meningioma with Tyrosine-rich crystals	1999	58-year diabetic female with 6-month history of headaches, black outs, dizziness	MRI: 4 cm dural mass in left parietal region	Fibrous meningioma	Interdigitating intermediate filaments with presence of osmiophilic crystals with petaloid appearance	Crystals seen were tyrosine rich
Schollenberg et al ^[7]	A Case of Clear Cell Meningioma with Tyrosine-rich Crystals	2012	54-year male with 2-year history of right lower limb and back pain	MRI: L3-L4 homogenously enhancing intradural mass and compressing nerve root	Clear cell meningioma	Interdigitating intermediate filaments with extracellular osmiophilic crystals with petaloid appearance and whorled collagen deposits	Crystals seen were tyrosine rich
Reinertsen et al ^[8]	Meningioma with Tyrosine-Rich Crystalloids: A Case report and review of the literature	2018	45-year-old female with long history of headaches, blurred vision and syncope	CT: Lobulated mass in right frontal lobe MRI: Extra axial dural mass from right orbital roof measuring 7.4x4.8cm. Significant mass effect present.	Atypical meningioma	–	Crystals seen were tyrosine rich

Table 1. Review of literature in chronological order

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