Watchful waiting for a Foramen Magnum Meningioma

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Introduction

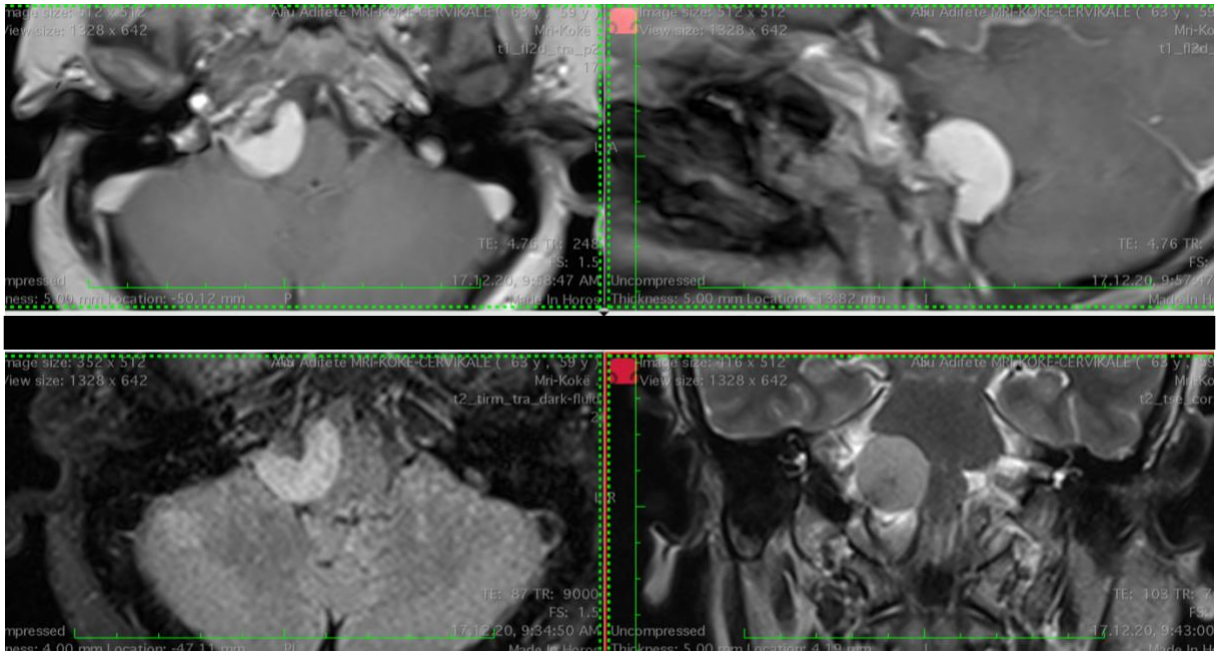
Meningiomas are the most common central nervous system tumor with an incidence rate of 8.14/100,000. Most meningiomas are benign, slow-growing, but with a consistent pattern of increasing incidence [2]. Meningiomas are classified based on morphologic criteria by the World Health Organization (WHO) into three groups (grade I-III) [3]. Tissue confirmation may be deferred in some patients, as meningiomas can be diagnosed by imaging [4,5], and are frequently diagnosed incidentally. In those cases, magnetic resonance imaging may be used to differentiate benign and atypical meningioma, based on tumor margins, edema, bone destruction, and diffusion coefficient [6,7]. Population-based studies estimate that around 90% are grade I (benign), 10% are grade II (borderline), and less than 3% are grade III (malignant) [1,8].

Foramen magnum meningiomas (FMM) are very rare and present formidable surgical challenge and carry the risk of postoperative complications because of their proximity to critical neurovascular structures.[1,2] Consequently, FM meningiomas are associated with a higher incidence of morbidity and mortality compared with meningiomas occurring in other locations. 2,3Given these factors, it is crucial to delve deeper into the molecular characteristics of FM meningiomas to advance our understanding of their pathogenesis and guide the development of effective therapeutic strategies.

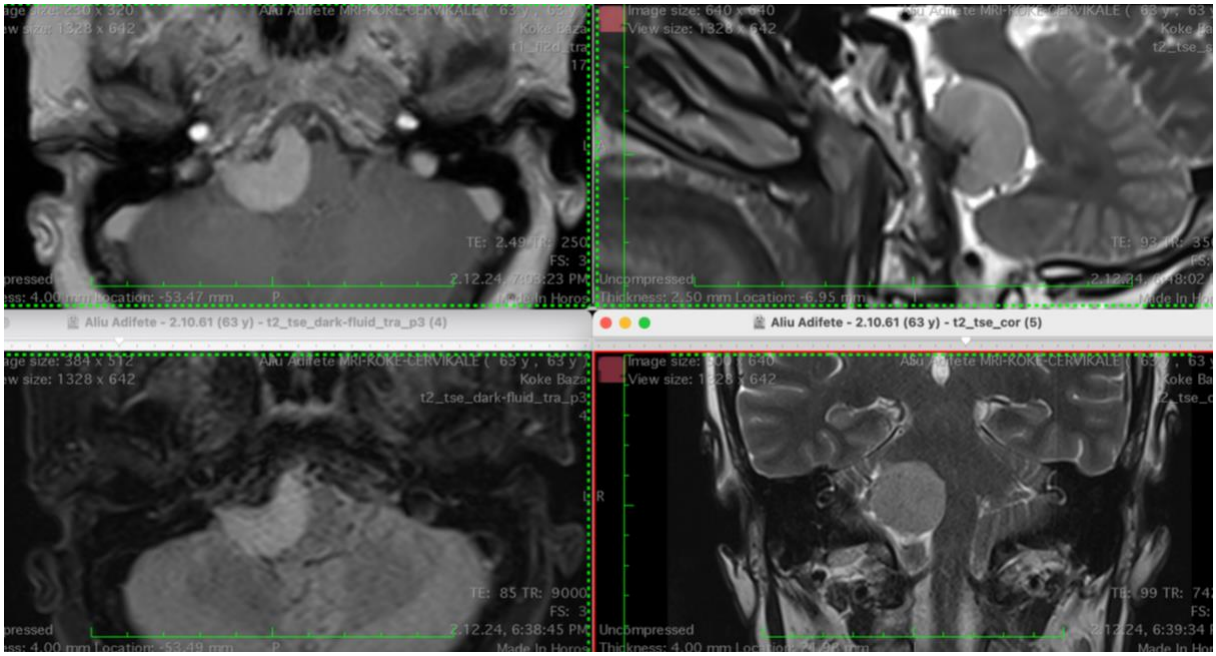
Treatment approaches for meningiomas include either observation alone, radiation alone, or surgical resection with or without radiation, and rarely, in cases with progression, a trial of systemic treatment, although there are no FDA approved therapies.

Case presentation

We present the case of a 68 y.o woman diagnosed five years ago with an incidental cranio-cervical junction lesion, with image characteristics of a Foramen Magnum Meningioma.
The patient reported no history of complains and on evaluation was neurologically intact, examined separately by two neurologists.
Laryngeal endoscopy was requested for evaluation of the vocal cords, which resulted normal.
MRI every six months was requested in the first year and then every year, which showed, no changes in size, consistence or perilesional edema/intensity.
In our series of 20 wait-and-watch Meningiomas according to the EANO guidelines, this is the only with a FM location.



MRI in 2020.



MRI in 2024

Discussion

According to the European Association of Neuro-Oncology (EANO) guidelines regarding the Meningioma management, combining data on patient age, performance status, co-morbidities, and MRI features (meningioma hyperintensity, peritumoral edema, proximity to neurovascular structures, size) patients are categorized as low, medium, or high risk for growth and progression, and an individualized monitoring strategy can be developed.

The incidence of clinically silent meningiomas in 75-year-old individuals was determined five years ago in the Vienna Trans-danube Ageing Study (VITA).
The conclusion drawn at the time was to recommend watchful waiting and perform imaging procedures regularly in the event of incidental, clinically silent meningiomas in elderly persons. Watchful waiting increased from 35.2% in 2004 to 51.4% in 2014 according to the American National Cancer Database.

In our case series of 32 FMM, all treated surgically, with outcomes, technical pearls and surgical approaches, published twice in EANS annual scientific meetings[16,17], this case is the only one which was managed through a watchful waiting approach.

In A population-based matched cohort study regarding surgically treated asymptomatic meningioma patients: In 36% of patients, preoperative occupation was not resumed, mostly due to cognitive symptoms.

The first description of the mutational profile specific to FM meningiomas, including TRAF7, AKT1, NF2, POLR2A, and KLF4 was published in July 2024.
Clinically significant driver mutations were detected in 58 patients (93.5%). TRAF7 (26, 41.9%) emerged as the most frequently mutated gene, followed by AKT1E17K (19, 30.6%), NF2 (11, 17.7%), POLR2A (8, 12.9%; 6 POLR2AQ403K and 2 POLR2AH439_L440del), KLF4K409Q (7, 11.3%), and PIK3CA (4, 6.5%; 2 PIK3CAH1047R and 2 PIK3CAE545K).

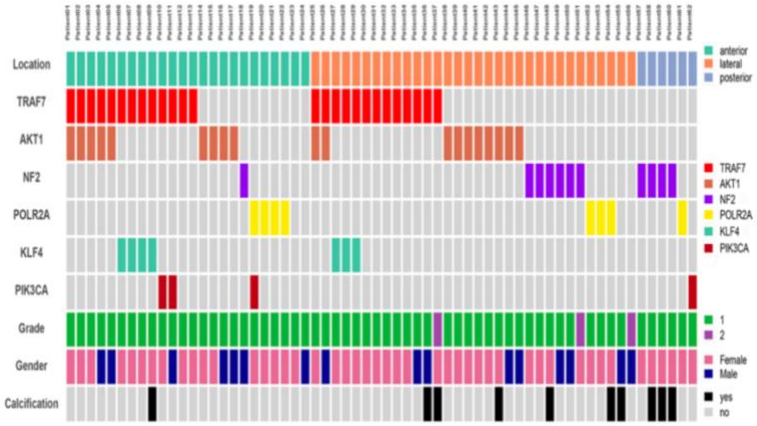


FIG. 1. Summary of clinical features and molecular alterations of 62 FM meningiomas. Figure is available in color online only.

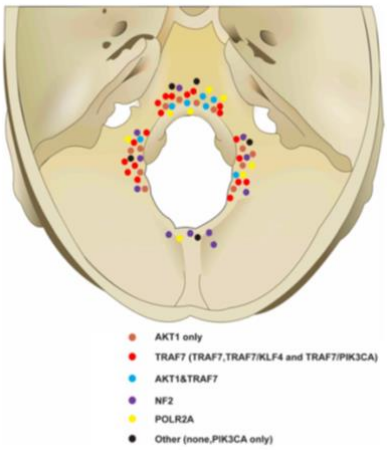
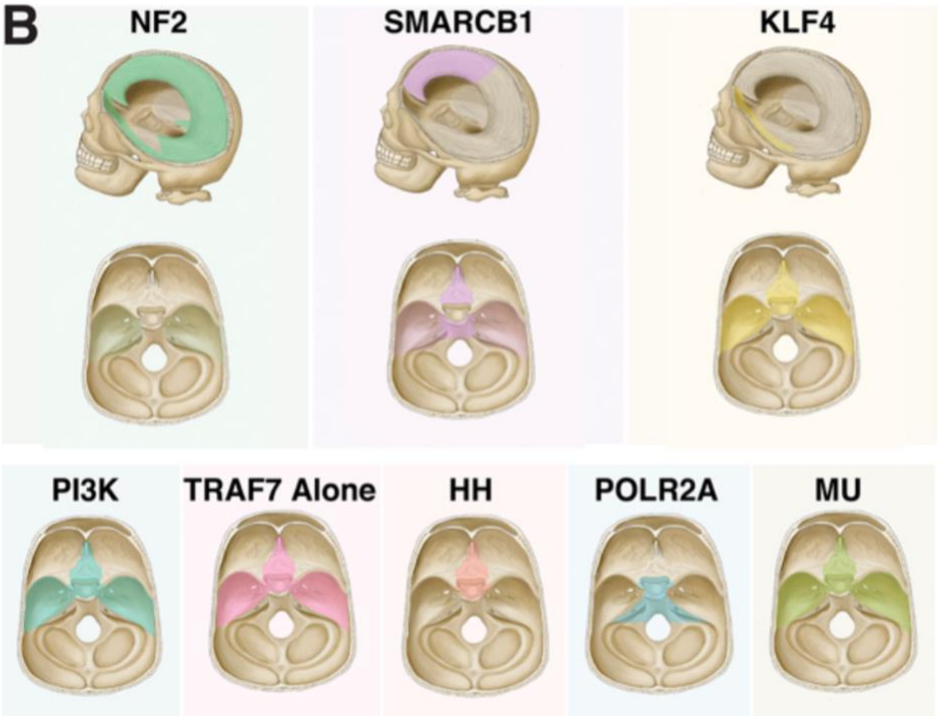


FIG. 2. Anatomical distribution of meningiomas along the FM and in relation to the brainstem. Figure is available in color online only.

All patients with POLR2A mutations were significantly younger at the time of first diagnosis and exhibited larger tumor volume at diagnosis compared with patients with NF2 or TRAF7 mutations who were older and harbored smaller tumors.
This suggests potential age-related differences in the development or progression of meningiomas associated with these specific mutations.

According to Moliterno et al. in the cohort study of 3000 Meningiomas, correlations between genomic subgroup and clinical features were seen also in relation to the location.
POLR2A mutants, predominant in the FM location, exhibited relatively less edema compared to other subgroups aside from SMARCB1.



Conclusion

The watch-and-wait approach recommended after several studies for incidental Meningiomas, considering also the molecular profiling and characteristics of the FMM, was confirmed by the present investigation and follow up of the case. No tumor growth was seen; no clinical symptoms have been registered thus far. The intervals between control investigations may even be prolonged depending on the location of the lesion.

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