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Trismus Masquerading Embryonal Rhabdomyosarcoma- an extremely rare case report and literature review

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Abstract: Rhabdomyosarcoma was first described by Weber[1] in 1854. It is the most common soft tissue sarcoma and malignant orbital neoplasm in infants and children[1, 2]. It is a rare malignant tumour with extremely aggressive and infiltrative nature[3], histologically;embryonal or botryoid type is the most common variant occurring in the head and neck region4. The alveolar type of rhabdomyosarcoma is mostly seen in extremities and trunk in children and young adults[2]. Here we present an extremely rare case of an embryonalrhabdomyosarcoma located in the maxillary sinus which extends into the orbital cavity with absolutely no ocular symptoms.

Key words:*Rhabdomyosarcoma; embryonalrhabdomyosarcoma; orbital rhabdomyosarcoma*.

I. Case Report:

An 11 year old apparently healthy female patient reported to the Department of Oral& Maxillofacial Surgery, Government Dental College, Kottayam, with a two month old history of difficulty and pain during mouth opening, which was started with a throat pain. There was no history of any kind of trauma or any history of any medical or surgical problems.

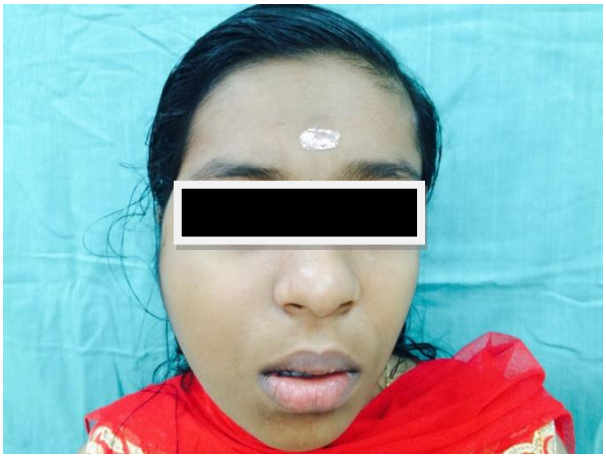


Fig 1:- profile view

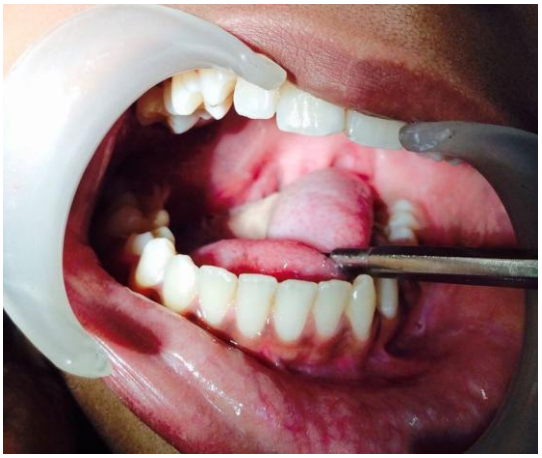


Fig 2:- intra oral

Clinical examination revealed a slight asymmetry of face on the right side and there was absolutely no odontogenic cause. Mouth opening was painful; interincisal distance was 20mm.There were no signs of nasal obstruction, motor disturbanceof the eyeball, or sensory abnormality of the trigeminal nerve.

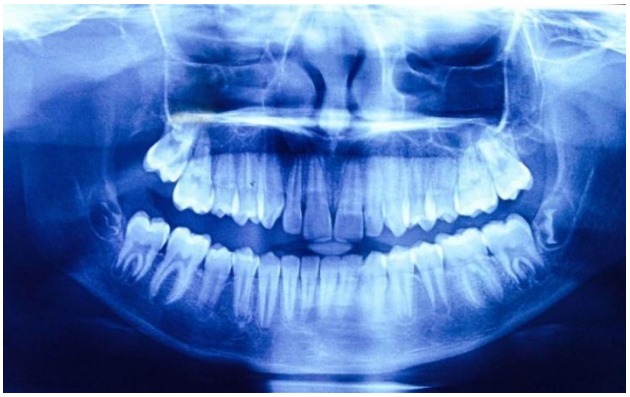
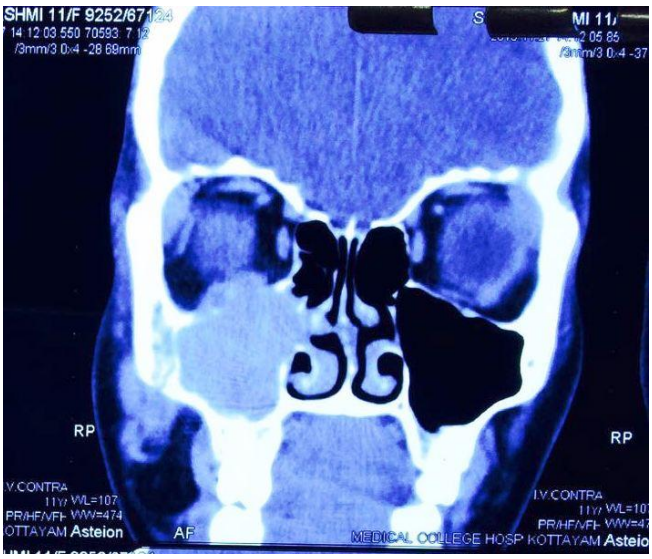


Fig 3:- OPG

A computed tomography(CT) and cone beam CT scan revealed an irregular heterogenoussoft-tissuedensity measuring 8cm x 6.5 cm x 6 cm noted in the right parapharyngeal space extending medially to the pharyngeal mucosal space thus obliterating the nasopharynx and oropharynx extending to the right maxillary sinus eroding its lateral wall, laterally to the infratemporal fossa and masticator space involving all the muscles of mastication, inferiorly up to a level 5mm above the hyoid bone.

Anterosuperiorly it extends into the inferior aspect of the extra corneal space of the right orbit by eroding the orbital floor and by eroding the base of the skull in the right side greater wing of sphenoid and temporal bone. Lesion in the right anterior temporal lobe measures around 2.5cm x 2.3 cm x 2cm. medially it is seen invading the right cavernous sinus also.



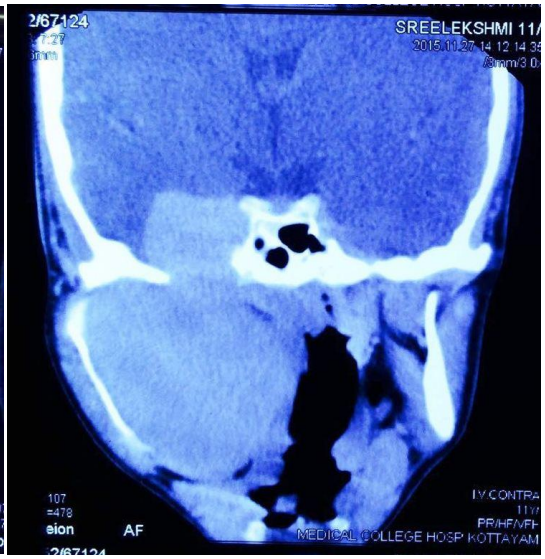
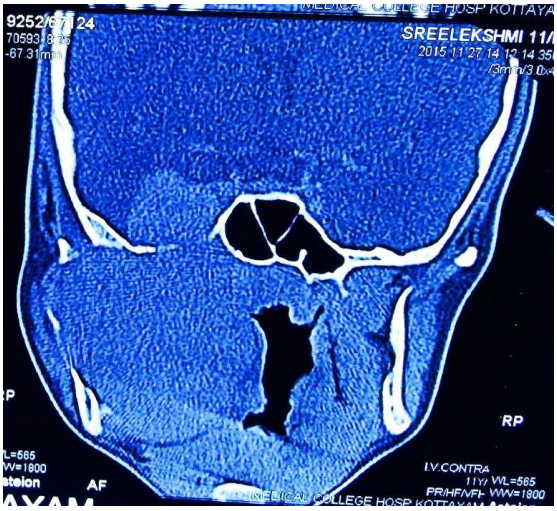


Fig 4:- CT coronal sectionFig 5:- CT coronal section

Posteriorly lesion is seen extending to the pterigopalatine fossa and the medial pterigoid plate is eroded. Lateraly the lesion is seen eroding the right ramus, coronoid and condylar processes of mandible and right zygomatic arch. Lesion is compressing and displacing the right side jugular vein and carotid artery posterolaterally.



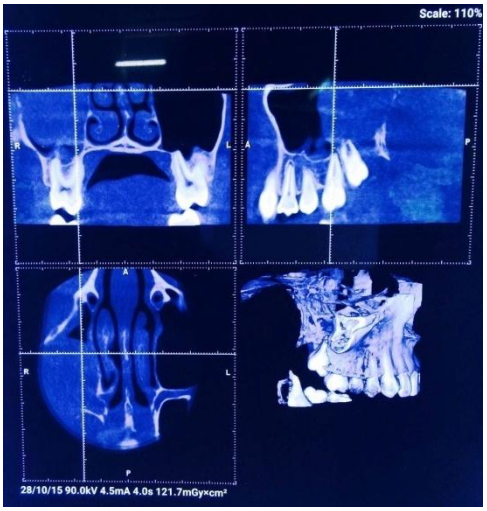
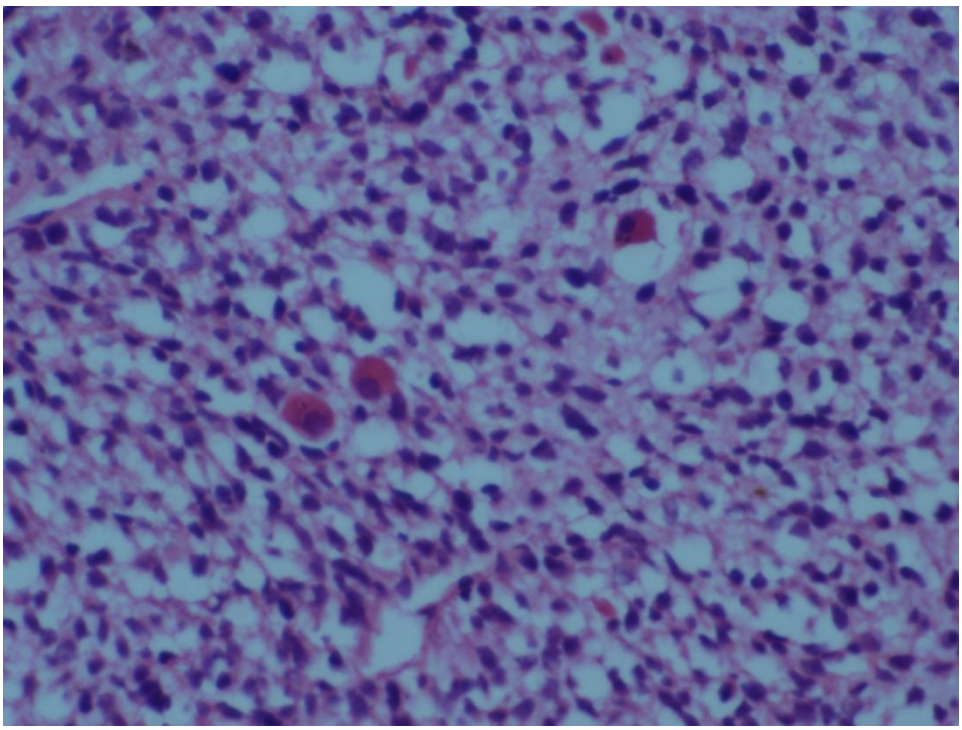


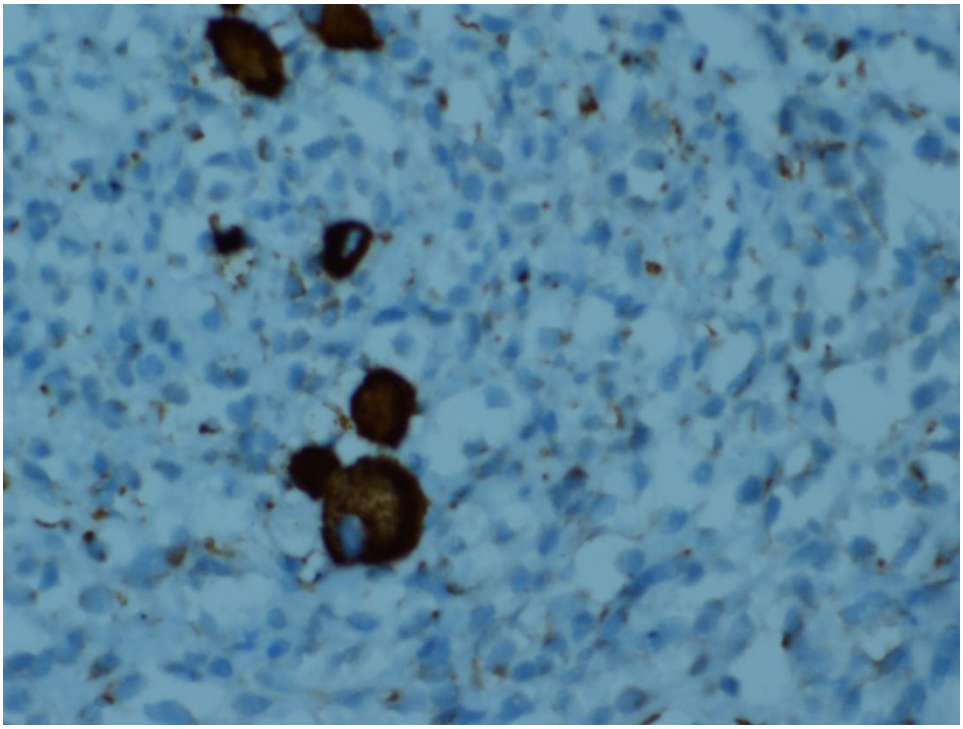
Fig 6 :- CT coronal section Fig 7:- cone beam CT

USG neck and abdomen was done and there were no signs of lymphadenopathy and hepatosplenomegally. With these we came to a differential diagnosis of minor salivary gland tumour, neurogenic tumour or rhabdomyosarcoma; an incisional biopsy was done intraorally but no conclusive finding was obtained and hence the patient was sent to our ENT unit. With the help of ENT unit, a sinus endoscopic surgery was done and a small piece of the fragile mass filling the right maxillary sinus was taken for histologic examination.

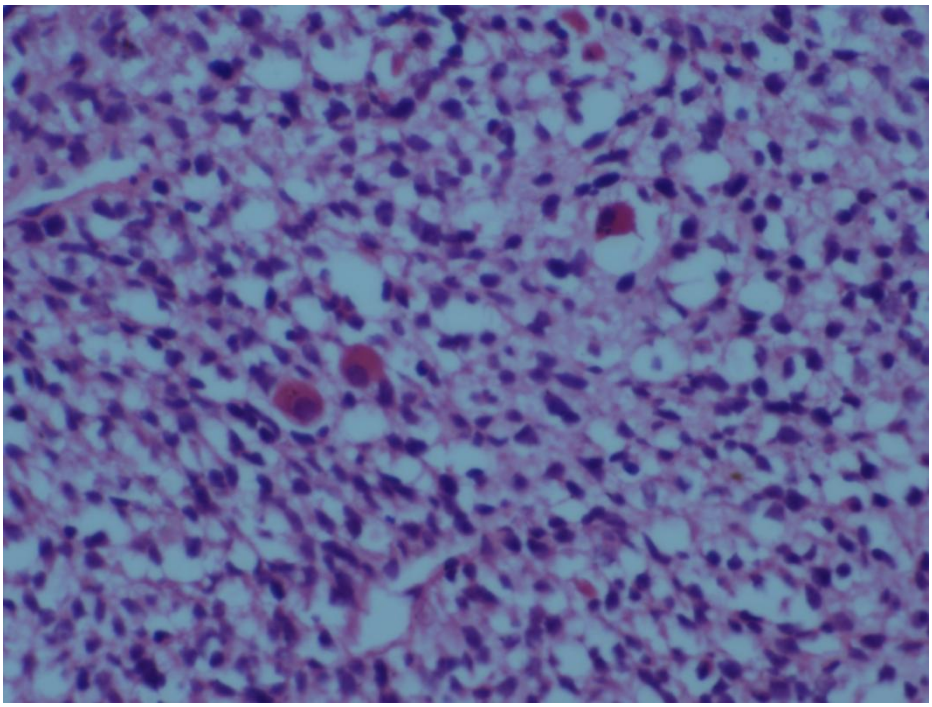
Microscopic examination of the biopsy section shows portion of skeletal muscle tissue with an infiltrating cellular neoplasm composed of sheets of spindly cells. Predominant cells are small round or spindly with eosinophillic cytoplasm and hyperchromatic nuclei, some of them strap like, Admixed with there are oval or spindly or tadpole shaped rhabdomyosarcoma cells,many rhabdomyoblasts seen, In Skeletal muscle tissue shows varying degrees of differenciation. Stroma shows myxoid and congested vessels.



Sheets of round to ovoid tumor cells with scattered rhabdomyoblasts having abundant esinophilic cytoplasm and eccentric nuclei.H & E 40 X



Desmin positivity in round to ovoid tumor cells and rhabdomyoblasts.IHC 40 X



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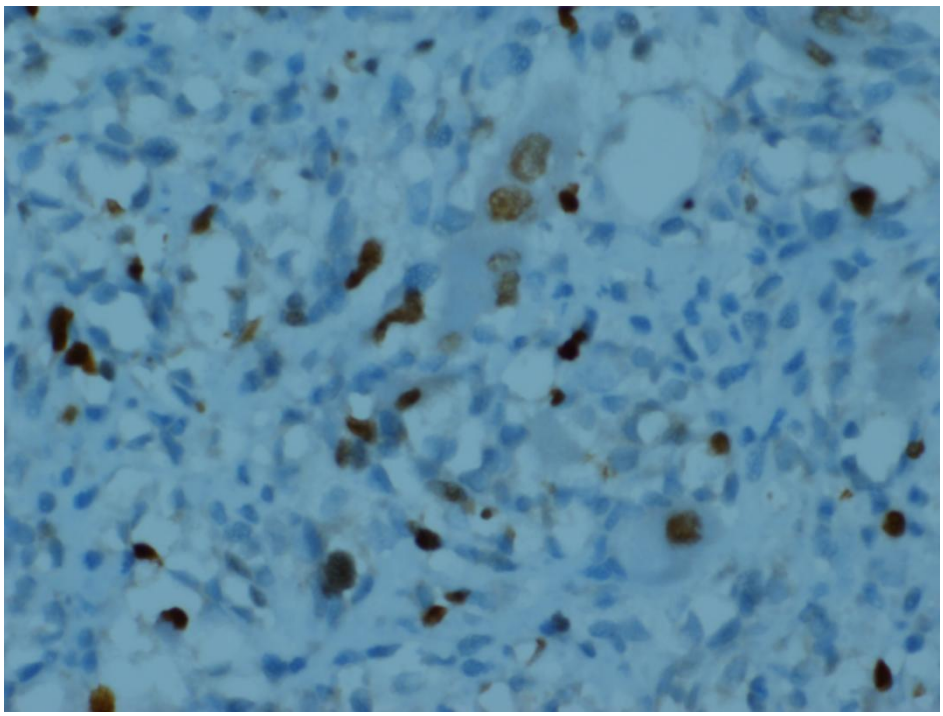

Myogenin positivity in round to ovoid tumor cells and rhabdomyoblasts.
IHC 40X

Immunohistochemically, mostof the tumor cells showed a positive reactionfor Desmin and some of them were alsopositive for myogenin and are negative for S100 protein. On the basis of radiographic, microscopic and immunohisochemical data, this lesion exhibits all characteristic features of spindle cell neoplasm with Rhabdomyoblastic features. Hence final diagnosis of embryonalrhabdomyosarcoma-spindle cell variant was made.

II. Discussion

Rhabdomyosarcoma is the most common childhood soft-tissue sarcoma of the orbit[5], Constituting 10% of all rhabdomyosarcomas[6]. Earlier it was thought to be arising from striated muscle cells; but immunohistochemistry has demonstrated that the tumor arises from pluripotentialmesenchymal cells that undergo malignant transformations[7]. 75% of orbitalrhabdomyosarcomas occur within the first decade of life and has a bimodal age distribution[7].

Due to the masquerading nature of the clinical presentationit was very difficult to establish a clinical diagnosis. In our case the symptom was difficulty and pain in mouth opening. The most important prognostic factors of rhabdomyosarcoma of orbit are the site, histologic features and disease dissemination. Theembryonal histology, and nonmetastaticnature of the disease also proved to be good prognostic indicators[8,9].

In 1997, the Inter RhabdomyosarcomaStudy group(IRS) published a report representing patients who were treated under protocols from trail I to IV[6], and categorized them under four subsets of orbital rhabdomyosarcoma. Tumors were catagorized as group I in 3% of patients,group II in 20%, group III in 74%, and group IV in 3% of patients.80% of them were embryonal in subtype, alveolar in 9%, botryoid in 4%, and anaplastic in0%. These figures are accepted by other large studies also[10].

Post operative chemotherapy and radiotherapy are recommended in majority of group III cases as the tumours are incompletely resected. In case of group I and Group II tumours are confined to the orbit in the initial stages[7].

Radiotherapy has complications to ocular structures as lens is extremelysensitive to irradiation and hence years later 90% of these can result in cataract. Orbital hypoplasia, keratopathy, dry eye,optic neuropathyandretinopathy are the major complications of high dose radiotherapy[5].These ill-effects can be masked by administering low dose radiotherapy with the use of shields and better localization[1].

Over the past few decades there has been a revolutionary progress in the availability of therapeutic options for orbital diseases. Complete resection of tumour, disfiguring enucleation and exenteration was the standard of care for rhabdomyosarcomaupto the 1970's. Because of the survival rate was just 35% after exenteration[5]. Radiotherapy followed by chemotherapy was administered in patients with recent or disseminated disease in the beginning of mid 1960's.

Based on tumour grade and stage; protocols combining radiotherapy and specific chemotherapeutic regimens were introduced in the 1970's by IRS. These treatment protocols are extremely specific and dynamic, which evolved over the course of various trials. The survival rate of patients with rhabdomyosarcoma dramatically increased to 93%[11] with incomplete excision of orbital tumour coupled with post operative radiotherapy and chemotherapy.

Our case ofSpindle cell Rhabdomyosarcoma with right infratemporal fossa, parameningeal with intracranial extension, IRS (International Rhabdomyosarcoma Society), Group 3, Stage III. The Child was sent to higher center for expert management and started on chemotherapy (Vincristine , Actinomycin-D& Cyclophosphamide) along with upfront radiotherapy in view of intracranial extension of the tumour.Metastatic work up including chest X-ray,bone marrow biopsy and cerebrospinal fluid study was negative.

Patients with rhabdomyosarcoma ofembryonaltype have more favourable prognostic result than those with alveolar or pleomorphic rhabdomyosarcoma. multiagent chemotherapymust be continued for 1-2 years[2].

III. Conclusion

Other than odontogenic and other Squamous cell neoplasms, Rhabdomyosarcoma is also should be a possible tumour in the oral and maxillofacial region. The tumour should be treated with multimodal therapy due to its aggressiveness and infiltrative nature. It is customary to submit all excised tissue for histopathologic analysis. This case report underscores the importance of the above tradition. Though lesions like Rhabdomyosarcoma are the exception rather than the norm, it is becoming of a prudent clinician to be on the lookout for such rare entities.

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SURGICAL PATHOLOGY SLIDE REVIEW REPORT

Biopsy

NATURE OF MATERIALS RECIEVED
Slide and block review

GROSS DESCRIPTION
Received 1 H&E slide and 1 paraffin block labelled 12232/15.

MICROSCOPIC DESCRIPTION
Section shows a cellular neoplasm composed of sheets of spindly cells having eosinophilic cytoplasm and hyperchromatic nculei.
Many rhabdomyoblasts seen, having abundant eosinophilic cytoplasm and hyperchromatic nuclei, some of them strap like.
The neoplastic ells are positive for desmin and myogenin and are negative for S100 protein

DIAGNOSIS
Rhabdomyosarcoma, spindle cell variant.

T Code C499 M Code 89103

Reported By:DR. THARA SOMANATHAN

Bone marrow bx.
Section shows spicules of bone c intervening marrow spaces showing erythroid, myeloid & megakaryocytic elements.
Bone marrow [illegible]
Cellular marrow showing erythroid, myeloid & megakaryocytic elements.

Slides and blocks will be issued 24 hrs after receiving written request from treating physician in RCC
Surgical specimens will be discarded after one month of despatch of reports.

Life Beyond Cancer