

Answers to Audience Questions - WSPOS World Wide Webinars

WWW 19 – Season 2 – Malignancies And The Paediatric Eye



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1. Maybe the protocols are different in Europe? I believed was not necessary to remove ALL Rabdo and survive rate was not better doing this.

Ap.R: In my clinical practice, I try to remove the entire tumor whenever possible.

NC: I agree, in France the management includes a biopsy, systemic chemo and focal proton beam.

SK: In our experience, RMS prognosis is better when there is a combination of excisional biopsy, chemotherapy and radiotherapy.

JP: As I've indicated during the discussion, the combination of chemotherapy and radiation, without relating to full excision, has made the difference in the good results of treating rhabdomyosarcoma, which is far better than several decades ago.

GS: I believe in safe debulking (avoiding muscle / globe damage) with the aim of minimizing total tumor volume/burden, which will help with hopefully reduced field / dose.

2. Is there gamma knife for orbit?

Ap.R: I am not aware of gamma knife use for the orbit.

NC: if proton beam is not available, stereotaxic radiotherapy can be used

JP: We use gamma knife for intraocular tumors and have no experience using it for orbital tumors. However, I do not see why it cannot be used.

GS: We do not use Gamma Knife Surgery for orbital tumors in general, as it delivers a very high dose in a single fraction (hence called Radiosurgery), to protect the globe, optic nerve and other sensitive structures. Hence fractionated doses.

3. What does everyone on the panel use for pharmacological confirmation of Horner syndrome?

As.R: Apraclonidine 0.5% if there is evidence of ptosis or other concerns (change in behaviour). Do not give just for anisocoria in small babies as physiological anisocoria is so common.

Ap.R: I use topical apraclonidine for children over 6 months

JP: I do not treat Horner's syndrome in our ocular oncology service

GS: Believe in the standard protocol. Apraclonidine is not readily available in several countries, now lately Cocaine also not available.

AE: Apraclonidine 0.5% even in young infants under the age of 6 months, with proper counselling regarding potential side effects and adequate observation time

4. I have a 3-month-old patient treated for mediastinal neuroblastoma with CBH syndrome. Is there treatment?

Ap.R: The ptosis can be surgically corrected after treatment of the neuroblastoma. Many cases of Horner's resolve after treatment of the primary cause.

JP: I have no experience.

GS: For patients with mediastinal/cervical Neuroblastoma a PET-CT along with Bone marrow biopsy (trephination, not aspiration). If genetics of tumors are negative for Nmyc, deletion at 10 & 11 q, patient is at low risk. Hence in the absence of metastasis and complete resection, surgery is usually curative. If metastasis present and genetics are negative, chemotherapy is recommended (intermediate risk protocol). If genetics show N myc amplification of 11 q, with or without metastasis, chemotherapy should follow High-risk protocol (inputs from Dr Miriam Kimpo, Pediatric Oncologist, NUH, Singapore)

5. Does an MRI definitely show carotid dissection? or do we need BT angio or MR angio?

As.R: I would always ask for MR angio in addition to MRI; this is very rare in children

Ap.R: I believe a CT angiogram is the most sensitive to diagnose dissection but I would discuss the case with the radiologist to determine the test and the earliest availability.

JP: I have no experience.

GS: CT angio is generally more sensitive and specific and helps direct management as well.

AE: MRI may not show the dissection. I usually discuss with the radiologist and primary team to decide on best approach which may be MRA or CTA depending on the general condition, ability to sedate or stay still for the scan, and suspected pathology.

6. What would be the treatment for multiple papilloma in addition to excision to prevent recurrence?

As.R: Use cimetidine or ranitidine for conjunctival papillomata.

Ap.R: For recurrent multiple papilloma I treat them with oral cimetidine.

NC: I used interferon eye drops but to date interferon alfa is no more available in Europe. MSD pharmaceuticals stopped the production. I have replaced interferon by FU eye drops only in multiple recurrences.

JP: We use interferon, but others have good experience with cimetidine

GS: Oral cimetidine where available. Practice no-touch technique during excision, use 10% iodine, occasionally even 95% alcohol to amputated base followed by saline lavages. First time is the best time.

7. What is the best ttt for dermolipoma involving the cornea?

As.R: Excision

Ap.R: I perform excision with amniotic membrane transplantation

NC: No treatments if visual acuity is not threatened. In case of involvement of the optic axis, an OCT of the cornea can help to manage the lamellar resection

JP: Dermolipoma is usually seen in the temporal periphery, and dermoid is the lesion seen in the limbus with involvement of the cornea. We usually only excise it. Of course, if the corneal center is involved, corneal graft may be considered.

GS: If significantly elevated, safe debulking avoiding corneal perforation esp. with high astigmatism. Lateral canthal reconstruction may be performed if malformed.

AE: Excision. If it is large with posterior extension, consider debulking and a 2 step procedure. First one would be debulking and separating the posterior aspect from the anterior corneal aspect. This allows easier and more controlled excision of the anterior part, possibly even its shrinkage somewhat. Then be prepared for a corneal graft if needed, May be lamellar. If very superficial may consider SLET.

8. How often do you examine children with conjunctival naevi? What is the follow up period?

As.R: 1 year

Ap.R: Conjunctival nevi rarely are malignant in children. I photo document and monitor annually.

NC: because there is a risk (rare) of melanoma arising from nevi, the follow up period is for life. The more important is education of the patient about the necessity of quick appointment in case of rapid increase of the lesion.

JP: Since conjunctival nevi in children rarely transform to malignancy, annual follow up is probably good enough. Unless, worrisome signs are present, then in the beginning once every 6 months.

GS: If significantly elevated, safe debulking avoiding corneal perforation esp. with high astigmatism. Lateral canthal reconstruction may be performed if malformed.

AE: Proper documentation with Photography, slit lamp photos, or even basic but accurate illustration in the medical record. Then start with 6 month follow and gradually go to yearly with its stability

9. Regarding RB early detection, my personnel experience is that leukocoria is too late sign / symptom. Is there any other screening modalities to pick up RB early in children, may be a community program?

As.R: If there is a family history, screening from birth under anaesthetic every month is recommended. This picks up smaller tumour. Also remember, Strabismus is a cause.

Ap.R: The only other option is dilated fundus exams. Considering how rare retinoblastoma is, it is not feasible to perform dilated exams on all children. Often times, there is a lag time between detection of leucocoria and medical consultation. By raising awareness, this lag time can be decreased.

NC: The best way of screening is dilated fundus that should be done in any case of leucocoria or strabismus. There is no rational to do a fundus in all the babies if we consider the incidence of Rb. For familial retinoblastoma however systematic screening of the fundus is mandatory. After birth then once a months until you get the genetic analysis. The child even if negative for Rb mutation, need to be screen regularly in case of mosaicism.

JP: Since RB is rare (about one in 15,00 live birth) there is no routine follow up during pregnancy (unless there is a family history). In Israel all new-born babies have red reflex examination before discharge, but not more than that.

GS: in our experience, strabismus has been an earlier sign. Leukocoria is a relatively late sign. Flash photography during birthday parties have also been an early sign in some of our patients. All pediatric ophthalmic exam for any reason < 5 yrs. of age should have a complete and dilated fundus exam.

AE: Any patient seen for the first time gets a dilated fundus exam and then at least once yearly even if following up for other reasons regardless of risk factors. Family history and other risk factors would then decide if other patients need more regular or thorough screening, according to guidelines

10. If a mass in the caruncle looking fleshy, can we just observe?

As.R: I will need to see a photo before I can say anything.

Ap.R: Malignant lesions of the caruncle in children are very rare. The only fleshy tumor in the caruncle that I have seen are pyogenic granuloma which can be observed but often require excision.

JP: I would remove **fleshy** caruncular mass, just to be on the safe side, although almost sure it will be a benign lesion. If just a small lesion, mostly nevi, follow up is enough.

GS: If asymptomatic and long standing yes. If symptomatic and more recent origin, can be safely excised.