

## **Summary of ECD Global Alliance Internet Chat**

### **20 Sep 2014**

#### 5 Attendees

- A member started by saying that she thought that there would probably be only a few Chatters because of the patient meeting. She was right, but those who did come on Chatted for quite a while. The member who had fallen and broken a rib was asked about. His wife told us that his rib has healed.
- The members who had not gone to the meeting are both having to travel a lot for treatment, etc. One has been having CHOP chemotherapy [(C)yclophosphamide, (H)ydroxydaunorubicin, (O)ncovin (vincristine), (P)rednisone], because she has recently been diagnosed as having a lymphoma, as well as ECD. This was diagnosed following a recent biopsy. The treatment is going well, although the doctors are having some trouble trying to mix the two treatments (for lymphoma and ECD). She was taking Kineret (Anakinra) and Methotrexate for the ECD (she has rheumatoid arthritis too, and methotrexate is used to treat that as well). She is having the CHOP chemo at present and hopes to be adding back the Anakinra soon. She is BRAF positive, but was getting good results while using Anakinra, so Dr. Janku was staying with this treatment.
- Another member wishes that she could have been able to go on to Vemurafenib (V). She has blurred vision and has lost her peripheral vision. First, it only affected the left eye, but now both are affected. She does not have any bulging. She also has macular oedema, and has had Avastin injections in the eye for that. She has no problem with bone pain. Her main complaint is with her vision. Her last PET-CT showed great improvement. She was asked whether Anakinra had been particularly effective on certain areas, and it was said that it was very helpful on long bones and some other areas.
- A member came on who is just starting Anakinra. Her story is yet another complicated one! She was diagnosed in 2012 with Rosai-Dorfman Disease, but the docs now say that it is actually ECD. She is BRAF negative, and is being treated with Anakinra. Her disease is in her orbits, long bones, and the left kidney. She has just started the Anakinra, and has only had 10 shots. She said that if she didn't have the disease in her orbits she wouldn't know that she had the disease. She went to Sloan for the first time in June, and then for her 2nd biopsy in July. She will probably have to get retested in 3 months. The first biopsy report said that the situation was "complicated", but Dr. McClain said it favored RDD. The NIH took a look at the pathology, and also said that it favored RDD. But a 2nd biopsy at Sloan favored ECD. The biopsies were performed on the front of her skull and on her tibia. Hopefully, there will be no more biopsies for a while, but she will be having a PET-CT, MRI, and CAT scan, etc.
- Another member has a similar pattern of disease. It affects his orbits, brain, joints, and long bones. He also has some double vision. His eyes were bulging and very pink. Since he started V, the pink eyes are gone. The tumors in the orbit have shrunk a bit, but are still significant.

When he was at Mayo, the ophthalmologist (with 30+ years of experience) was telling him that he had never seen optic nerves swollen as badly as his! He was almost going to start treatment with cladribine (2CDA), but then the idea of going to Sloan-Kettering came up. It does mean a lot of effort traveling monthly, with overnight stays every other month. The V is "working fine", but the scans tell him that he is not out of the woods yet. He has "a lot of side effects" from the V and, at times, he needs to take breaks. He was also diagnosed with lymphoma after his first MRI, and then the doctors seemed to go along with that as the only diagnosis. He was diagnosed as having ECD in May this year, but thinks that he must have had it for at least 5-10 years. He does not see an ophthalmologist at Sloan, but goes back to Mayo for his eyes. He believes that if the orbital tumors can be controlled, then his eyes would be OK. He told us that he had to have surgery, so that a biopsy could be taken from behind his left eye. This "was a very painful experience". The surgery was just to get the biopsy, but the doctor injected some steroid into his eyes and this helped control the inflammation tremendously.