

Summary of ECD Global Alliance Internet Chat

4 Feb 2017

11 Attendees

- It was a well-attended Chat. One or two of those who signed in did not actually post anything.
- At first, a chatter told us that she was at a gathering "but pulled away for a few minutes to borrow the local Starbucks wifi!". Very sensible! She has just had a baseline echo, and this week she has her first blood draws since beginning dabrafenib (D). The echo was OK, she thinks. At least no phone calls from the doctors! She has a few lesions in her cerebellum and brainstem. Those on her legs have gone away; a few outside her heart and on her sternum seem to be "sleeping". When she was on vemurafenib (V) the ones in her brain had not been affected, and she also had had bad joint issues, so she changed from V to D in January. She has noticed some back pains, but doesn't know whether D has that as a side-effect.
- A member had bad pains in his back. On the previous night, he had got out of bed and then fallen. His wife had gotten up to make sure that he was OK. His hips are "so bad" that he gets thrown off balance 2-5 times a week.
- A member who has not been on the Chat for a while filled us in on his story. He is taking cobimetinib (C) medicine and is in the third phase of treatment. He goes to Dana Farber in Boston and sees Dr. Eric Jacobson. [Dr. Diamond also prescribes Cobi to some patients as part of a Cobi trial at Memorial Sloan Kettering Cancer Center (MSKCCA).] He was diagnosed last February and was initially on interferon, but with no results. Since then his symptoms are much the same, and the pains that he gets seems to be spiking higher again. The cobimetinib is showing no change in the tumor size.

His team has decided to continue on with the C for another two months, and then do a body scan with a brain MRI. At that time, they may have to make a change if there is no difference.

He said that he has continuous head pain from a tumor growing throughout his skull that is expanding the skull thickness. That is where he believes that the bone pain is coming from. He thinks that he has ECD that is not typical in that he has tumors in his head, neck, and spine, and also has other more widespread ECD lesions.

He has been taking Tramadol and oxycodone when the pain goes beyond level 5 static pain on a pain score. This only reduces the pain back to a level 2. Pain cycles last up to 40 hours. The only thing that helps him is to lean forward, and to keep his head low. His body doesn't hurt like his head, even though he has tumor all the way down his spine. His doctors are wondering whether some of the tumors could be of a different type, and that is why they are not seeing results just yet. His last scan showed that the ECD had spread now to his left side.
- Another member said that he has a lot of pain in the legs, but also in the back, the fingers, and the neck. But the pain is not always in the same place!
- Another member on C came on. She has started her 8th cycle. She used to see Dr. Jacobsen in Boston, but now sees Dr. Diamond in NYC. She has a mix of Rosai Dorfman Syndrome (another very rare histiocytic disease) and ECD. Her pain is in her bones, which is hard to treat. The activity level of her tumors has calmed down.
 - Edema (or Oedema to Europeans) was discussed. (<http://www.mayoclinic.org/diseases-conditions/edema/basics/definition/con-20033037>) This is fluid that collects in the tissues, so that they are swollen. Most cases affect the ankles, and you can make impressive

dents by pushing down on the swelling. This may be due to the ECD, or other prescribed medication. Also, dietary changes can bring it on. Finally, heart and kidney failure can cause it as well.

Sitting with your feet up on a footstool may help, but your feet have to be HIGHER than your bottom, or nothing will happen! The doctors can often help with a diuretic drug, this will make you urinate more often.

- Diabetes was discussed. Diabetes insipidus is a problem caused by failure of the anterior-pituitary gland in the brain to produce anti-diuretic hormones, (nothing to do with sugar). This results in you passing massive amounts of weak urine and being very thirsty. It is very common in ECD and treated with desmopressin tablets or nasal spray.
- Diabetes mellitus is the "sugar diabetes". It is the production of too much urine, but this is due to there being too much sugar in the blood. This passes out through the kidneys and "pulls" with it water by osmosis, leaving the patient dehydrated. This is treated with insulin injections or tablets.
- A member in remission is going to have another brain CT. His wife, and others, are finding his speech more slurry.
- A member's case history has been presented in Park City, Utah at a Winter Skin Seminar. She wasn't asked to show up, but they used her records of the last few years. It was a dermatology conference. For ten years she had been told that her skin lesions were "something else"! She is pleased to be able to help educate dermatologists to look further, and not just stop when they think that they have found a box to put us in.